Aus der

Klinik für Herz-. Thorax- und Gefäßchirurgie

Deutsches Herzzentrum Berlin

Arztlicher Direktor: Prof. Dr. med. Dr. h.c. mult. Roland Hetzer

Habilitationsschrift

SURGICAL TECHNIQUES AND OPERATIVE OUTCOME OF REPAIR OF MITRAL VALVE LESIONS IN INFANTS AND CHILDREN

zur Erlangung der Lehrbefähigung

für das Fach Herzchirurgie

vorgelegt dem Fakultätsrat der Medizinischen Fakultät

Charite-Universitätsmedizin Berlin

von

Dr.med. Eva Maria Delmo Walter

Berlin 2012

Eingereicht: August 2012

Dekanin: Prof. Dr. med. Annette Grüters-Kieslich

1. Gutachter: Prof. Dr. med. Dr. h.c. Friedhelm Beyersdorf
2. Gutachter: Prof. Dr. med. Dr. h.c. Christian Schlensak

To my son, Mariano

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Abbreviations

ALCAPA	Anomalous origin of the left coronary artery from the pulmonary artery
ALRP	Anterior leaflet retention plasty
ASD	Atrial septal defect
AV	Atrioventricular
CAVSD	Complete atrioventricular septal defect
CHD	Congenital heart disease
IMI	Ischemic mitral incompetence
LVOT	Left ventricular outflow tract
LVOTO	Left ventricular outflow tract obstruction
MV	Mitral valve
MI	Mitral valve insufficiency
MR	Mitral regurgitation
MS	Mitral valve stenosis
NYHA	New York Heart Association
SAM	systolic anterior motion
TEE	Transesophageal echocardiography

1.0 Introduction

1.1 Background

Mitral valve (MV) disease in infants and children is rare.[1,2,3,4] Abnormalities in MV structure and function in this population may either be congenital or acquired lesions.

Congenital malformations of the mitral valve are complex, typically associated with other congenital heart diseases, and are rarely isolated.[5,6,7] Abnormalities in MV anatomy and function are commonly seen in common anomalies [ventricular septal defect (VSD), patent ductus arteriosus, aortic stenosis, coarctation of the aorta,[8] tetralogy of Fallot, transposition of the great arteries, and truncus arteriosus] and in several complex congenital heart malformations [atrioventricular septal defects (AVSD), endocardial cushion defect, abnormal atrioventricular connections, and hypoplastic heart syndrome and occasionally associated with heterotaxy].[1,5,8,9,10,11] These associated congenital heart lesions may hide, or be hidden by, the MV malformations. In addition, several syndromes are associated with abnormalities in MV anatomy or function, including connective tissue disorders (Marfan's and Ehlers-Danlos syndromes),[9,12,13,14,15,16,17] Shone's anomaly,[10,18,19,20] endocardiofibrosis, hypertrophic cardiomyopathy. It may also be involved in congenital polyvalvular dysplasia (Bharati-Lev disease),[21] congenital mucopolysaccharidosis,[22] especially Hurler's syndrome,[23] and may lead to severe MV malfunction in young children.

Acquired MV lesions in infancy and childhood are infectious, rheumatic and degenerative. Acquired mitral insufficiency occurs with anomalous left coronary artery (Bland-White-Garland syndrome) and fibroelastosis, caused by papillary muscle infarction.[24]

A clear understanding of the surgically relevant anatomy and morphology of the mitral valve is a key to a successful establishment of sound indications for surgery and is essential to successful repair and the avoidance of injury to the aortic valve, conduction system, and coronary vessels (Fig. 1).

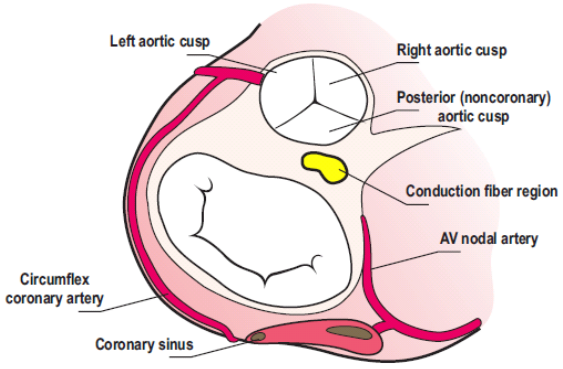


Fig. 1. Surgically relevant mitral valve anatomy

1.2 Surgical morphology of the mitral valve

The mitral valve complex [25] is made up of several individual parts, which need to function in harmony if the overall complex is to maintain its competency. This consists of the annulus, commissures, leaflets, the tendinous cords, and the papillary muscles (Fig. 2). The *mitral annulus* is an integral part of the fibrous skeleton of the heart. It separates the atrium and the ventricle and coincides with the annulus of the aortic valve beneath the left coronary and non-coronary aortic cusps, abuts the tricuspid annulus at the membranous interventricular septum, and lies deep within the left atrioventricular groove beneath the coronary sinus and left circumflex coronary artery. The relationship of the annulus to the atrioventricular groove and its contents, the circumflex coronary artery, requires constant awareness to prevent injury to the artery during MV surgery. It is a dynamic structure and functions as a sphincter in systole, producing an elliptical orifice with a cirumference of approximately 8 cm.[26,27,28] The normal MV has two *leaflets*, anterior and posterior. The larger, wider, anterior leaflet attaches 150° to the annulus, and is trapezoidal in shape. As a consequence of being in fibrous continuity with the aortic valve, it forms the boundary of the left ventricular outflow tract. Two ends of this fibrous continuity anchor the aortic-mitral valve unit across the short axis

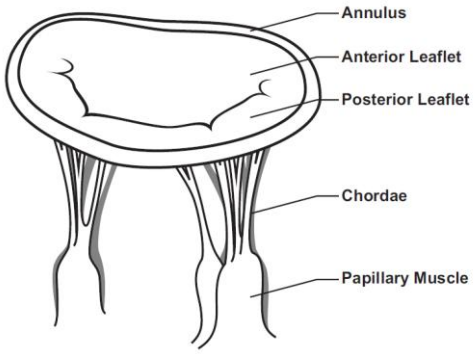


Fig. 2. Mitral valve complex

of the left ventricle, where it becomes thickened to form the right and left trigones.[29] The posterior leaflet is longer and narrower and occupies 210° of the annulus. These leaflets are flexible, have smooth surfaces, and are largely composed of collagenous tissues. The posterior leaflet is often scalloped, with a large central scallop flanked by two smaller (commissural) scallops [30] (Fig.3). The middle scallop is the largest in over 90% of valves studied [30] and is the portion most often affected by chordal rupture. The posterior leaflet occupies 65% of the annular circumference, but the surface areas of both anterior and posterior leaflets are nearly equal (4.9 cm ^{2} versus 5.0 cm ^{2} respectively).[30] This combined area (9.9 cm^2) is nearly twice the area of the systolic annular orifice (5.2 cm^2). This is because the valve does not naturally close at the leaflet-free margins, rather a large area of leaflet tissue overlaps with the opposing leaflet, thereby forming a tight seal. This segment of leaflet tissue between the free edge and the line of valve closure is termed the *rough zone*, characterized by

thickened nodular ridges. Interdigitation of these nodular ridges enhances sealing of the valve. The portion of the leaflet extending from the line of valve closure to the annulus is referred to as the *clear zone* and is noticeably thinner and smoother (Fig. 4). The demarcation between rough and clear zones identifies the normal line of valve closure.[30] The two leaflets are separated by the anterolateral and posteromedial *commissures*. Although commissures do not extend all the way to the annulus, they can be consistently identified by the insertion of a single, fan-shaped chord arising from the tip of the adjacent papillary muscle. Beneath the commisures lies the two corresponding *papillary muscles*, which are extensions of the subendocardial myocardium. The two papillary muscles are separate, distinct structures which are attached only at their bases to the ventricular wall. The posterior muscle is attached to the junction of the interventricular septum and posterior left ventricular wall and is supplied by branches of the posterior descending coronary artery.[31] This papillary muscle holds chordae connected to both leaflets around the posterior commissure. The anterolateral papillary muscle, supplied by branches of the circumflex marginal coronary artery, is attached to the lateral ventricular wall and controls chordae near the anterior commissure. Thus, each papillary muscle anchors chordae from both leaflets. The free edges of the two leaflets merge with thin, string-like second-and third-order chordae which fuse together to form four-to six-first order chordae at the tip of each papillary muscle. Chordae tendinae arising from the papillary muscles insert on both sides of the corresponding commissures; hence each leaflet is supported to the free edge and rough zone by chordae from both papillary muscles and by basal chordae directly attaching to the wall of the left ventricle.[29] Commissural chordae rarely elongate; hence can be used a a reference point for proper closing plane of the valve.

During systole, the proper length, position, and function of the papillary muscles and chordae arrest the upward motion of the two leaflets at the level necessary to achieve anatomic closure of the mitral valve. During diastole, blood reaches the left ventricle by passing between the two leaflets and through the interstices of the chordal network.[32]

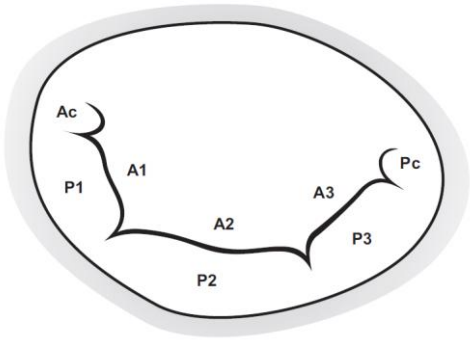


Fig. 3. Surgically relevant segments of anterior and posterior leaflets

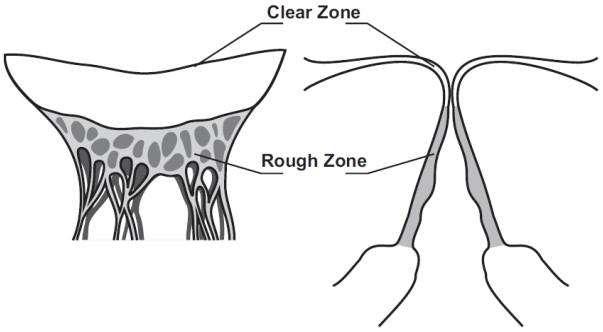


Fig. 4. Coaptation surfaces of the mitral valve complex

1.3 Pathologic anatomy of the mitral valve

Functionally, MV anomalies can be primarily insufficient, stenotic, or mixed lesions. Structurally, any of the five components of the valve complex can be abnormal or missing. The degree of structural abnormality varies from mild to severe with variable number of affected components. Although vivid descriptive terms such as parachute, hammock or floppy valves are helpful, the pathologic anatomy is most easily understood by describing observed abnormalities of the different valve components.[5,7,11]

1.4 Pathophysiological classification of mitral valve disease

Transthoracic and transesophageal echocardiography allows classification of the MV malformations according to its pathophysiology and according to function.[6] The malformations, classified according to its pathophysiology, are described as mitral valve incompetence or mitral valve stenosis, whether the motion of the leaflets is normal, prolapsed or restricted and whether or not there are abnormal papillary muscles (Table 1). When more than one anomaly of the mitral valve apparatus is present, the primary and predominant lesion serves to classify the malformation. *This pathophysiological classification has been proven to be an important guideline in the application of surgical techniques of valve repair*, the primary objective of which is to restore the normal valve function rather than a normal mitral valve anatomy.

1.5 Functional classification of mitral valve disease

The malformations are further classified by the recognition of the most characteristic feature, as seen from the left atrial view during surgery. Carpentier's functional classification of mitral insufficiency[33,34,35] is clearly described in Table 2.

Table 1. Pathophysiological classification of mitral valve disease

Mitral valve insufficiency[33]	**Mitral valve stenosis**[11]	
Type I ***Normal leaflet motion*** The regurgitation results from lack of leaflet coaptation Annulus dilatation Cleft leaflet Partial leaflet agenesis	**Type A** ***Normal papillary muscle*** Commissural fusion Supravalvar ring Annulus hypoplasia Excess valvar tissue Obstructive left superior vena cavas	**Type I** ***Typical congenital mitral stenosis*** Short chordae tendinae Obliteration of interchordal spaces by fibrous tissue Variable reduction of interpapillary distance
Type II ***Leaflet prolapse*** The free edge of one or two leaflets overrides the plane of the orifice during systole Chordae and/or papillary muscle elongation Absence of chordae	**Type B** ***Abnormal papillary muscles*** Parachute valve Hammock valve Absent papillary muscle	**Type II Hypoplastic congenital mitral stenosis** Almost always associated with hypoplastic heart syndrome
Type III ***Restricted leaflet motion*** The motion of one or the two leaflets is limited. *With normal papillary muscles* Commissural fusion Short chordae *With abnormal papillary muscles* Parachute valve Hammock valve Papillary muscle hypolasia or agenesis		**Type III** ***Supramitral ring*** With or without other types of congenital mitral stenosis **Type IV Parachute mitral valve**

1.6 Segmental classification of mitral valve disease[6]

Segmental classification by transesophageal echocardiography allows precise location of the leaflet dysfunction. Likewise, in addition to valve analysis, echocardiography allows assessment of the volumes, ventricular contractility and recognition of associated lesions. However, it is rarely possible to make a precise diagnosis of all the lesions responsible for the MV dysfunction by echocardiography. The precise diagnosis is always made **intraoperatively**.

Table 2. Functional classification of mitral valve disease

Classification	Lesions	
	Congenital malformations	Acquired lesions
Type I Normal leaflet motion The regurgitation results from a lack of coaptation between the leaflets.	Annulus dilatation Leaflet defect Cleft leaflet Partial leaflet agenesis	Annulus dilatation Leaflet perforation
Type II Leaflet prolapse The free edge of one or of the two leaflets overrides the plane of the orifice during systole.	Chordae agenesis Chordal elongation/rupture Papillary muscle elongation/rupture	Chordal rupture Chordal elongation Papillary muscle elongation
Type III Restricted leaflet motion. The motion of one of the leaflets is limited. This leads to mitral valve stenosis; however, valvular insufficiency may occur with certain lesions. Two subgroups are described, depending upon the morphology of the papillary muscles.	A.***With normal papillary muscle*** Papillary muscle commissure fusion Leaflet thickening Short /thickened/fused chordae Ebstein-type mitral valve Excessive leaflet tissue Valvar ring Annular hypoplasia B***. With abnormal papillary muscles*** Parachute valve Hammock valve Absent papillary muscles hypoplasia or agenesis	Papillary muscle rupture Due to valvular lesions Commissure fusion Leaflet fusion Calcification Due to subvalvular lesions Short chordae Fused chordae

1.7 Congenital mitral valve lesions

The congenital anomaly may involve any component of the mitral apparatus and may result in stenosis, with or without regurgitation, or in pure regurgitation. Although one component may be involved, more often the entire valve is affected.

1.7.1 Congenital mitral insufficiency (MI) may result from annular dilatation secondary to anterior or posterior leaflet prolapse or to posterior leaflet hypoplasia with chordal shortening. Chordal elongation and valve prolapse may be so severe that chordal rupture can develop, even in young children, producing severe regurgitation. Congenital mitral regurgitation may also be produced by clefts or gaps, or perforations in the anterior mitral leaflet, accessory commissures, or leaflet hypoplasia at medial or lateral commissures.

Classified as ***Type I MV insufficiency*** (Fig. 5a) by Carpentier, [33] the mitral valve has normal leaflet motion, but insufficient because of either inadequate central coaptation on the basis of *annular dilatation and deformation.* This may occur primarily without any associated lesions. Carpentier and colleagues[7] found essentially isolated annular dilatation in 8 (17%) of 47 cases with congenital MV disease, although some deficiency of commissural tissue is implied in their description. In some cases annular dilatation is secondary to mitral insufficiency from some other valve deformity. The dilatation may affect the posterior leaflet, which may be slightly thickened. The basic valvar anomaly leading to insufficiency may be subtle and difficult to identify. *Cleft leaflet* also belongs to this category. This condition usually affects the anterior leaflet. It may be of two types: cleft anterior leaflet or cleft in the atrioventricular septal defect.[36] In cleft anterior leaflet, the MV apparatus is normal, with the exception of a cleft that vertically separates the anterior leaflet into two hemi-leaflets.[37,38,39] The cleft is directed towards the right and non-coronary interleaflet triangle. The two papillary muscles are normal and normally located, with the two chordae connected to the respective aortic fibrous trigone. The commissures are correctly positioned, and the posterior leaflet attachment represents two-thirds of the circumference of the mitral orifice, as in normal hearts. Abnormal chordae tendinae may be present and are often attached to the free edges of the leaflet. Cleft posterior leaflet is rare [33] and is often confused with enlargement of the normal indentations of the posterior leaflet, secondary to annular dilatation. These indentations are normal features of the posterior leaflet.[30] Only clefts that affect the leaflet tissue between these indentations may be regarded as *true clefts*. Cleft may simply represent leaflet deficiency in the area without chordal support.[40,41] *Leaflet defect*, such as holes [42] or localized agenesis of the leaflet tissue,[43] may be encountered particularly in the posterior leaflet. The free edge of the defect may be free of chordae or may have thin chordae tendinae attachment. The combination of a young age of the child and absence of any history of bacterial endocarditis are evidences of the congenital nature of these lesions.

The second classification ***Type II*** (Fig. 5B)**,** allows regurgitation of blood because of ***leaflet prolapse,*** a condition in which the free edge of one or two leaflets overrides the plane of the orifice during systole.[7] *Absence of chordae* [44] in the margin or a whole leaflet segment likewise results in insufficiency. *Chordal elongation* usually involves all of the chordae tendinae arising from one papillary muscle. The corresponding portion of the leaflet tissue is prolapsed within the atrium during systole.

In the third classification, ***Type III A*** (Fig. 5c), the valve has ***restricted leaflet motion and normal papillary muscles.***[33] *Commissural fusion* is seen with a tissue obliterating the commissural area covered by an abnormal endothelial tissue, sometimes multiperforated, without the characteristic fibrosis and calcifications seen in rheumatic valve disease. Underneath the fused commissures, chordae and hypertrophied papillary muscles may be seen adherent. MV insufficiency results from either hypoplasia or a defect of the anterior leaflet or annular dilatation, or both. *Short chordae* are seen well-delineated and only slightly thickened without the fusion seen in rheumatic heart diseases. The interchordal spaces are reduced, which, in conjunction with papillary muscle hypertrophy, may create an associated subvalvar stenosis. The annulus is often dilated secondary to the mitral valve regurgitation.

In ***Type III B*** classification, the valve has ***restricted leaflet motion and abnormal papillary muscles.***[33] *Parachute mitral valve*, although purely stenotic, may be associated with insufficiency, due to anterior leaflet hypoplasia near one commissure. The *hammock valve,* as in parachute valve, is usually stenotic. Whenever an insufficiency is associated**,** it is due to leaflet hypoplasia or restricted leaflet motion. *Papillary muscle hypoplasia or agenesis* may lead to mitral valve incompetence. Chordae are attached to tiny papillary muscles or directly to various points of the ventricular wall. Valve regurgitation is caused by either abnormal tension on these chordae or underdevelopment of the leaflet tissue.

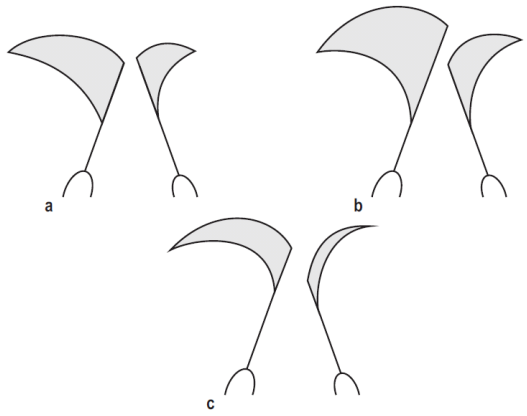


Fig. 5. Classification of malformations according to the motion of the leaflets and the morphology of the papillary muscles.[33]

1.7.2 Congenital mitral valve stenosis (MS) with or without regurgitation may result from supravalvar, valvar or valvar narrowing and may be accentuated by subvalvar obstruction from hypertrophied and misplaced papillary muscles or fused chordae. Frequently, stenosis is the result of abnormalities at multiple levels. Although embryologic origins of these complex anomalies are poorly understood, recent studies suggest that abnormal development of a transient left ventricular structure, a horseshoe-shaped ventricular myocardial ridge, results in various obstructive lesions, including parachute valve and formation of asymmetric valves.[45] Ruckmann and Van Praagh[11] have clearly classified congenital mitral stenosis (MS) into four types: ***Type I*** is the ***typical congenital mitral stenosis*** (Fig.6.1), in which the leaflet margins are rolled and thickened, the chordae tendinae are shortened and thickened, the interchordal spaces are partially or completely obliterated by fibrous tissue, the papillary muscles are underdeveloped and the interpapillary distance is mildly reduced. ***Type II*** is the ***hypoplastic MS*** (Fig.6.2), in which the MV orifice is small, the chordae tendinae are shortened but usually not thickened, with small papillary muscles and the interpapillary distance is maintained. This type of MV abnormality is distinguished from the normal MV such that all components of the valve are in miniature. This form of MS is associated with severe left ventricular outflow tract abnormalities. ***Type III***, the ***supramitral ring***,[5,10,46,47] (Fig.6.3) is a circumferential ridge of fibroconnective tissue originating at the atrial surfaces of the MV leaflets and causes various degrees of MV inlet obstruction. The ring is truly supravalvar at the level of the posterior leaflet (2-3 mm above the annulus), but intravalvar at the level of the anterior leaflet to which it is attached (3-5 mm below the annulus). Depending on the diameter of the ring, various degrees of obstruction may exist. This abnormality is usually associated with other anomalies of the MV apparatus, especially parachute mitral valve, a hammock valve and typical congenital mitral stenosis. ***Type IV*** is the ***parachute mitral valve*** (Fig.6.4), one of the most common malformations causing MV stenosis. All the chordae tendinae insert into a single papillary muscle. The chordae tendinae are usually shortened and thickened, the mobility of the leaflets is reduced and the effective MV orifice is decreased. The single papillary muscle is formed either from the fusion of the two papillary muscles or from the development of one of the papillary muscles, the second being either hypoplastic or adherent to the muscular ventricular wall without any chordal connection. The anterolateral papillary muscle is usually missing. The arrangement of papillary muscles, is variable [5,10,41,48,49,50,51] The stenosis results from obliteration of the interchordal spaces by excess valvar tissue. In some cases, a commissural cleft, usually anterior, corresponding to the absent papillary muscle, is the only orifice that allows blood to flow from the left atrium to the left ventricle. The presence of a parachute mitral valve does not automatically warrant a surgical indication. Some parachute mitral valves are functionally adequate. When the gradient through a restrictive parachute valve increases, one should suspect a supravalvar mitral ring.[52]

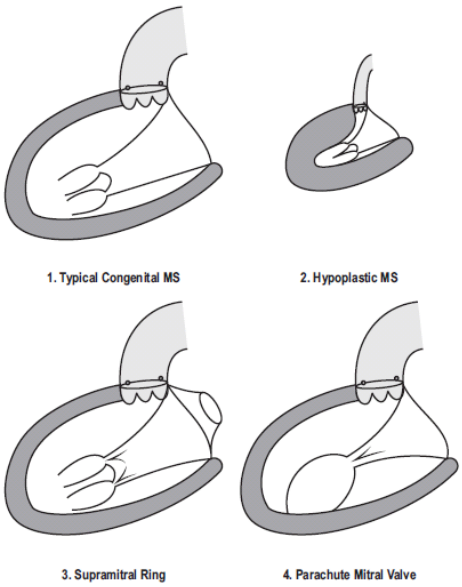


Fig. 6. Graphical illustration of Ruckmann and Van Praagh's [11] classification of congenital mitral stenosis

Other MV lesions causing MS, not identifiable in Ruckmann and Van Praagh's classification include ***congenital mitral valve stenosis with normal papillary muscles***. This condition arises from commisure fusion/short chordae, wherein one or two papillary muscles are directly implanted on the commissure without any intermediate chordae tendinae. The commissures are fused and thickened. The motion of the leaflets are restricted. The central orifice of the mitral valve between the two papillary muscles is severely narrowed, and the valve may have a funnel–shaped appearance.[41] A *forme fruste* may coexist, with short chordae instead of papillary commissure fusion. Another form associates one papillary commissure fusion and short chordae from the other papillary muscles. *Excess valvar tissue and double mitral orifice* may give rise to MS. In this condition, the interchordal spaces are obstructed by abnormal valvar tissue. The MV apparatus appears otherwise normal. In some cases, a large bridge of valvar tissue joins the anterior and posterior leaflets, thus delineating an accessory orifice of the mitral valve. *Annular hypoplasia*, an underdevelopment of the MV annulus can be associated with each of the previously-mentioned malformations. This must be clearly distinguished from MV hypoplasia and MV atresia, which constitute part of the hypoplastic left heart syndrome. The size of the mitral valve is compared with normal references for age and weight [53] (see Table 1, Original study, p.47).

Mitral valve stenosis with ***abnormal papillary muscles*** includes parachute mitral valve, which has been previously described. Belonging to this is *hammock valve*, which refers to the atrial aspect of various subvalvar anomalies.[54] The valvar orifice is partially obstructed by intermixed chordae and abnormal papillary muscles, characteristically implanted underneath the posterior leaflet. This malformation

includes various lesions such as mitral arcade, which has been described in literature as *mitral arcade, obstructive papillary muscles, hypertrophied papillary muscles,* and "typical" congenital stenosis.[5,50,55] These malformations actually follow a similar pattern: the two normal papillary muscles are absent, in their place are numerous papillary muscles and fibrous bands implanted high on the posterior wall of the ventricle underneath the posterior leaflet. As a result, there is lateral and upward displacement of the papillary muscles and obstruction of the MV orifice. The chordae tendinae of the anterior leaflet cross the orifice toward the posteriorly implanted papillary muscles, producing the hammock appearance. The leaflet tissue is often normal and pliable. There is not one central orifice but several orifices, in contrast to other types of MS In extreme cases, the hammock valve contains a fibrous diaphragm with scattered holes that allowed the blood to flow from the left atrium to the left ventricle. This malformation is the ***most difficult to correct by reconstructive techniques***. *Absent papillary muscles* rarely gives rise to mitral valve stenosis. Numerous intermixed chordae are attached to the ventricular wall with imperforate interchordal spaces.

1.8 Acquired mitral valve lesions

1.8.1 Infective endocarditis

Childhood infective endocarditis (IE) was previously only a rare complication of rheumatic heart disease.[56] However, the growing number of patients living with surgical repairs of congenital lesions now represent the majority of patients developing IE in childhood.[57] MV lesions in infective endocarditis may be manifested in various forms, i.e. annulus dilatation, commissural shortening, thickening of mitral valve leaflets, leaflet perforation, leaflet vegetation, leaflet prolapse, chordal elongation, chordal tears or rupture , and or combination of these lesions.

1.9 Statement of the Problem

With the aforementioned complexity of MV lesions, surgical management of mitral valve disease in infants and children has been always a major therapeutic challenge. It confounds special surgical difficulties because of the wide spectrum of morphological abnormalities requiring meticulous modifications of techniques of valve repair,[58,59,60] a high incidence of associated cardiac anomalies,[60,61] and relatively limited experience in each surgical center. Repair and reconstructive surgery of the mitral valve is a complex and technically demanding procedure requiring great experience of the surgeon. A thorough knowledge of the anatomy and function of the mitral apparatus, as well as the pathological processes involved in the lesion, is essential, since different techniques and modifications are necessary to repair a congenitally anomalous valve and to repair a diseased mitral valve from an acquired lesion. The surgeon, therefore, has to identify all abnormalities and plan the optimal way of achieving the maximal valvar competency in the operating room; hence, intraoperative decision-making and creativity are highly important. It is an enormously demanding

and formidable task for the surgeon to reconstruct and repair mitral valves in infants and children, primarily because of their size, the immature and fragile leaflets tissues in infants, as well as the associated congenital cardiac abnormalities, which take particular hemodynamic consideration. Mitral valve repair in children is guided by the same surgical rules as in adults, but the anatomic substrate differs greatly. The technical difficulties vary according to the anatomy, the size and age of the patient. The indications for surgery and the timing of surgery have to take into account a large range of issues and are therefore less straightforward than in adults. The first and foremost consideration when considering repair of the mitral valve in children is ***growth of the valve*** along with their somatic growth. Hence, the **principal issues in mitral valve repair in infants and children** are the *surgical techniques* employed according to the presenting morphology of the mitral valve, and the *modifications* necessary when there are other existing obstructive lesions, to obtain an optimal valve competency. The surgeons must establish *principles of mitral valve repair* and guidelines to evaluate the adequacy of the repair.

One major problem that confronts us is the *approach* in children with mitral valve lesions and with *small left ventricle*.

Another major issues are the operative results and the long-term functional outcome of the surgical techniques we employed in this group. These issues have been largely tackled at Deutsches Herzzentrum Berlin and are summarized in this dissertation from cumulative publications.

2.0 Surgical strategies

2.1 Echocardiographic evaluation

2.1.1 Mitral insufficiency is quantified by measurement of both the regurgitation volume and regurgitant jet.[9] At Deutsches Herzzentrum Berlin, using the color-doppler flow studies, quantification of mitral regurgitation is undertaken by measurement of the differences between mitral stroke and aortic stroke volume. The phenomenon of proximal isovelocity flow area, which corresponds to anatomic leakage, is used to measure the regurgitant volume know as proximal flow convergence. Regurgitation volume of <30 ml indicates *mild* regurgitation, 30-59 ml *moderate*, and >60 ml as *severe* regurgitation. Based on regurgitant fraction (%), severity of regurgitation is quantified as follows: *none*, 0%; *mild*, less than 20%; *moderate*, 20%-40%; *moderate to severe*, 40%-60%; and *severe*, exceeding 60%. In general, the proximal isovelocity area and distal jet area, which are both measures of the leakage area of the mitral valve in Doppler, virtually define regurgitation.

2.1.2 Mitral stenosis is quantified by measurement of the mitral valve orifice area (cm^2) and mean resting end-diastolic gradient (mmHg) and was graded from *0* (4-6 $cm^{2,}$ 0 mm Hg), *I-mild* (2-4 $cm^{2,}$ <5 mm Hg), *II-moderate* (1-2 $cm^{2,}$ 5-10 mm Hg), to *III -severe* (<1 cm^2, >10 mm Hg. Assessment of MV

function included: planimetric evaluation in mid-diastole of MV motion (leaflet mobility), determination of MV and orifice area; mitral flow is assessed using continuous wave-Doppler by measuring mean transmitral diastolic pressure gradient and systolic pulmonary artery pressure during the maximum velocity of tricuspid regurgitant flow; valve anatomy is evaluated as to valve thickness, commissural fusion, valve pliability, and morphology of the subvalvar apparatus.

2.2. Hemodynamic evaluation

At Deutsches Herzzentrum Berlin, a gradient ≥5 mm Hg across the mitral valve is considered significant to warrant intervention.

2.3 Principles of mitral valve repair

Hetzer R and **Delmo Walter EM**. Repair of congenital mitral valve insufficiency. Oper Tech Thorac Cardiovasc Surg: A Comparative Atlas 2010;15(4):260-272.

Hetzer R, **Delmo Walter EMB,** Huebler M, Alexi-Meskishvili V, Weng Y, Nagdyman N, Berger F. et.al. Modified surgical techniques and long term outcome of mitral valve reconstruction in 111 children. Ann Thorac Surg. 2008;86:604-613.

At Deustches Herzzentrum Berlin, we strongly believe that *repair for MV lesions allows undisturbed somatic and valve growth*, delays the need for future valve replacement and avoids anticoagulation. We assume that most, if not all, mitral valves repaired during childhood may eventually have to be replaced at some time in life. Thus, the concept of repair for MV lesions in childhood *is primarily aimed at growth* of the patient to an age when, if necessary, an adult-sized prosthesis can be implanted. This is best achieved by a spectrum of repair techniques applied individually without using any kind of prosthetic material. Repair is performed using only sutures and autologous pericardium. We use untreated autologous pericardium for posterior annulus reinforcement and as a pledget material. When used as an annular reinforcement, the pericardial strip should not lead to further annulus shortening; however, it should stabilize the suture-dependent repair and increase the height of the posterior leaflet coaptation.

In infants and children, the **primary goal** in MV repair is to achieve complete and rapid closure of the mitral orifice by a well-mobile anterior leaflet and a sufficiently large coaptation area, by bringing the posterior leaflet closer to the anterior leaflet. There should be no sutures placed along the anterior leaflet annulus. Depending on the particular etiology of the MV incompetence, several repair techniques may be required to achieve a competent valve. One must remember that the minimal final MV opening area is 3.5 cm^2 in adults and should not be smaller than 10% below the norm according to the body surface area in children.[55] Precautions must be observed to avoid the systolic anterior

motion (SAM) phenomenon caused by excessive annulus reduction and too wide a coaptation area, visible by "folding" of the anterior leaflet when the valve is tested with saline instillation into the ventricle.

Intravalvular saline injection and intraoperative esophageal echocardiography (TEE) are routinely performed to assess the adequacy of repair.

Regardless of the underlying pathological findings and techniques used, no patient leaves the operating room and be discharged from the hospital with more than mild mitral insufficiency or mild mitral stenosis.

2.4 Approach to mitral valve

Mitral valve repair is performed through either a median sternotomy which is optimal if other associated congenital heart lesions are present, especially in infants and young children, or a right anterolateral thoracotomy in the 5th intercostal space [62,63,64] which is a relatively good approach for exposure of the atrioventricular (AV) valves, especially when the patient has had previous surgery and a repeat median sternotomy would prove technically difficult due to scarring and adhesions; in adolescents and adults, it may be the patient's choice for cosmetic reasons. MV repair in infants and children is always performed under cardiopulmonary bypass, with aortic and bicaval cannulation, with the cavae snared. Before aortic and bicaval cannulation, the pericardium is opened longitudinally and a strip of pericardium is prepared free from adipose and extrapleural tissues, left unreated and set aside for later use. Antegrade intermittent cold crystalloid cardioplegia infusion every 10 to 15 minutes for myocardial protection is the rule.[65,66,67] Blood cardioplegia and normothermic bypass is used in older children and cold crystalloid cardioplegia in moderate hypothermia, (28°C to 32°C) is preferred in infants and small children for myocardial protection. Cardiac vent through the left atrium is employed to unload the left ventricle until discontinuation of extracorporeal circulation.

2.5 Exposure of the mitral valve

We expose the mitral valve and the subvalvular apparatus through a left atriotomy via a direct incision along the interatrial groove [62] Alternatively an approach via the right atrium and the interatrial septum (transseptal incision can be applied in cases of additional surgical steps on the right heart, preexisting defects of the septum or large ventricles[62] This incision is then carried through the septum longitudinally through the fossa ovalis, directed towards the entrance of the superior vena cava**.** This approach provides excellent exposure of the MV leaflets and the subvalvular apparatus,[62] in even the smallest atria, seen in infants and small children.

We usually prefer the left atriotomy approach. With a scalpel, a vertical incision is made along the interatrial groove,[62] and extended cephalad beneath the superior vena cava into the roof of the atrium and caudad under the inferior vena cava to the bottom of the left atrium. The heart is gently rotated with retractors to expose the mitral valve towards the surgeon.

2.6 Assessment of valve anatomy

We give particular attention to the leaflet coaptation, the chordae and position of the papillary muscles.[62] The leaflet coaptation is assessed with a forceful injection of saline with a bulb syringe through the valve. This maneuver demonstrates prolapse of any portion of the valve, if present. Traction sutures maybe placed at each commissure to elevate the annulus and allow better appreciation of the valve [62] To evaluate the leaflet apposition, a nerve hook is used to draw out the posterior leaflet near the anterolateral commissure, done similarly with the opposing portion of the anterior leaflet. There must be good apposition of the leaflet edges when sufficient tissue along a coaptation plane exists.[62] The coaptation of the anterior and posterior leaflets are assessed using the same nerve hook. We carefully take note of the sites of prolapse in a particular leaflet. In addition to leaflet and subvalvular apparatus, the size of the annulus is evaluated. Any significant dilatation will usually be obvious, as seen by the inability of the leaflets to coapt after transvalvular saline injection.[65]

2.7 Measurement of mitral valve diameter

It **i**s very important to measure the size the valve. We use either a special valve sizer called Ziemer-Hetzer valve sizer.[62] The nomogram published by Rowlatt et al.[53] is helpful in determining the normal valve diameter for a specific body surface area. We always make sure that the MV orifice area after repair is not smaller than 10% of the value specified in the nomogram [53] in order to prevent stenosis. When we are doubtful about the size of the valve after placement of the annular shortening sutures, the Ziemer-Hetzer valve sizer may be inserted through the valve and the sutures tied with the sizer in place. In this way, overtightening of the sutures which could induce stenosis is avoided.

2.8 Mitral valve repair techniques

Various repair techniques are used depending on the underlying MV pathology. Because malformations resulting in a particular MV lesion usually imply several anomalies, several repair steps may have to be used in the same patient. As a rule, we use the strongest available sutures which come with a comfortably small needle. The size of the needle is eventually the surgeon's preference. This takes into account that the enormous power of growth will eventually tear even strong sutures. The preferred sutures used for repair in infants are 5-0 polpropylene and of 3-0 polypropylene in

children. We always used untreated autologous pricardium as pledgets and as annular reinforcement strips of autologous pericardium.

2.8.1 Congenital mitral valve insufficiency

2.8.1.1 Annular dilatation in newborns and small infants

Annular dilatation in newborns and small infants is best repaired with a modified Kay-Wooler-Hetzer annuloplasty, modified by using pledgets of untreated autologous pericardium.[62] This is done by anchoring a double-ended polypropylene suture to an autologous pericardial pledget. A mattress suture is started by placing the needle through the fibrous body of the trigone, and both sutures are ran along the annulus fibrous tissue, approximately 1/4 of the posterior annulus length. After attaching a pericardial pledget, the sutures are firmly tied. The same procedure is performed on the opposite trigone.[62] This effects shortening of the posterior annulus. This is also the preferred technique in critical ischemic mitral incompetence along with repair of Bland White Garland syndrome.[62,63]

2.8.1.2 Severe annular dilatation in children and adolescents

Severely dilated annulus is best repaired with a modified Paneth-Hetzer technique.[62] This is performed by anchoring an autologous pericardial pledget to a 3-0 polypropylene suture with a relatively small needle. This suture is ran through the fibrous body of the trigone and tied. Then it is continued along the annulus from one trigone towards the middle section of the posterior annulus. The same is done on the opposite trigone. These sutures are then tied over an appropriately-sized Ziemer-Hetzer valve sizer in order to prevent over-narrowing of the valve orifice. The valve is then tested with saline injection for competence. Then using the same needles, both sutures are passed onto an untreated autologous pericardial strip.[62] Then the pericardial strip is attached or laid onto the posterior annulus from the midsegment towards the trigone using continous sutures anchored to the pericardial pledget and previousy-placed trigonal suture. These are then tied firmly with precaution to avoid any further narrowing of the orifice.[62] The same is done on the opposite side. We place a few additional interrupted sutures along the pericardial strip for reinforcement. Again, the nomogram of Rowlatt [53] is helpful in determining the normal valve diameter for a specific body surface area, to prevent stenosis. The leaflet coaptation is tested by a forceful transvalvular injection of saline with a bulb syringe to look for residual regurgitation. We also use this technique in ***anterior leaflet prolapse***, the then wider coaptation plane will eliminate prolapse. Shortening the posterior annulus produces wide and even coaptation, in such a way that when the anterior mitral leaflet closes, the border between the smooth and rough surface of the anterior leaflet forms the closure line with the posterior leaflet, without folding. We observed utmost precautions to avoid the systolic anterior motion (SAM) phenomenon seen as “folding” of the anterior leaflet when the valve is tested with

saline instillation into the ventricle.[62] The folding appears when the valve opening is made too narrow by overshortening (overreduction) of the posterior annulus.

2.8.1.3 Prolapse of posterior leaflet from ruptured chordae

Prolapse can occur anywhere along the posterior leaflet but is most commonly found in the region of P2. [62] This may lead to chordal rupture. We use the modified Gerbode-Hetzer plication plasty.[62] The flail segment is plicated towards the left ventricle in a V-shaped suture line of interrupted mattress suture of double-ended polypropylene sutures with pledgeted untreated autologous pericardium. Thus, the P1 segment is attached to the P3 segment**.** The sutures are then tied firmly**.** The valve is then tested for competence. When competence is assured, we check the valve diameter (size appropriate for BSA) as described in section 3.7. When competence and size are satisfactory a strip of autologous pericardium is sutured continuously onto the posterior annulus starting from the middle of the posterior annulus up to both trigones where the sutures are tied to the separate sutures which had been previously anchored onto the commissural fibrous body supported with pericardial pledgets. No further annular narrowing is effected by the pericardial strip. A few interrupted sutures are placed in addition to the continuous suture line along the autologous pericardial strip, particularly on both sides immediately next to the plication suture line at the level of the annulus .[62]

2.8.1.4 Mitral cleft

Differentiation between the cleft of an otherwise normal MV and the cleft seen in atrioventricular septal defects (AVSD) is important for its surgical indication and management.[65] The mitral cleft is assessed as to the presence of sufficient tissues .[62]. A transvalvular saline injection brings the cleft in apposition. This maneuver helps to expose the entire length of the cleft, from the fibrous skeleton to the chordae. Cleft maybe positioned centrally or asymmetrically or at any point on the posterior leaflet. The chordae are then inspected. When sufficient leaflet tissue is present on both sides of a cleft, we correct the cleft by suturing the edges of the leaflet together from where the cleft begins, up to the free margin of the leaflet with interrupted polypropylene suture.[62] The cleft should be completely closed and the valve opening diameter should be at least 10% below the norm based on the body surface area dependent-mitral valve diameter.[53] In clefts with an otherwise normal mitral valve, repair is generally accomplished by suturing the edges of the cleft, which should not induce mitral stenosis. *Direct and complete suture of the cleft is the rule*. It is our protocol to close the cleft whenever possible. However, if insufficient coaptation is achieved after cleft closure, a posterior annulus shortening, either modified Kay-Wooler or modified Paneth-Hetzer annuloplasty [62] may have to be added. Mitral cleft in our series is restricted to the anterior leaflet. The posterior leaflet appeared normal in size. We found neither accompanying annular dilatation nor abnormal subvalvular

apparatus or subaortic obstruction due to the chordal attachments of the cleft. The necessity of closing the mitral cleft is the most controversial issue in the surgical treatment of the mitral cleft [66] because of the tendency to cause stenosis.

This is different from our experience in atrioventricular septal defects, where we had cases in which the cleft could be closed only partially using an age-related minimal normal valve diameter as a guide to prevent valve stenosis, as was also reported by Alexi-Meskishvili from our group.[66,67]

2.8.1.5 Mitral valve insufficiency from lesions with restricted leaflet motion

Delmo Walter EM, Komoda T, Siniawski H, Hetzer R.Surgical reconstruction techniques for mitral valve insufficiency from lesions with restricted leaflet motion in infants and children. J Thorac Cardiovasc Surg. 2012,143:S48-S53.

Because congenital MI from restricted leaflet motion is clinically important although a rare entity, various repair techniques and results have been reported.[68-73] Approach and exposure of the mitral is described in sections 3.4, 3.5 and 3.6. The annulus, leaflets, chordae tendinae, and papillary muscles are exposed and meticulously inspected to determine the precise nature of the lesion, with particular attention given to the leaflet motion, position of the papillary muscles, and annulus. Leaflet apposition is assessed with a forceful injection of saline through the valve. The coaptation of the anterior and posterior leaflet with regards to the presence of sufficient tissues along the coaptation plane is assessed.[62] After confirming that leaflet coaptation is restricted giving rise to MI, the valve diameter is assessed as described in section 2.7.

Various repair techniques are employed according to the cause of restricted leaflet motion and presenting valve morphology.

Restricted leaflet motion with normal papillary muscles is brought about by fused commissures, thickened leaflets, short or matted chordae. In children with fused commissures, commisurotomy is done on both the anterolateral and posteromedial commissures. Division and splitting of papillary muscles are done in patients with short and matted chordae. In both aforementioned procedures, stabilization of posterior annulus with autologous pericardial strip [62,63] is performed.

Restricted leaflet motion with abnormal papillary muscles is due to hammock valves, parachute valves and papillary muscle hypoplasia. *Hammock valve* is dysplastic with shortened chordae directly inserted in a muscular mass of the posterior LV wall resulting in tethering of both leaflets, associated with annular dilatation, making the valve predominantly incompetent. In the absence of any papillary

muscle, a suitably thick part of the posterior left ventricular wall carrying the rudimentary chordae was carved off the wall.[63] Then it must be assured that both the remaining left ventricular wall and the "new papillary muscles" maintain sufficient muscle thickness to carry out their.[63]

Parachute valve has the usual two MV leaflets and commissures, but all the chordae tendinae cluster into one major papillary muscle. It presents as a funnel-type structure with some distinct fibrous lines at the sites of commissural fusion. The most appropriate site for leaflet-splitting incisions is defined on both sides from the common papillary muscles towards the "assumed" trigones.[63] These incisions are extended into the body of the papillary muscle which is then split towards its base assuring sufficient thickness of both new "papillary muscle heads".[63]

The degree and extent of incision, commissurotomy and fenestration are determined by measurement with a valve sizer evaluated according to the minimal age-related acceptable MV diameter, to avoid mitral stenosis. In both parachute and hammock valves, posterior annuloplasty reinforced with untreated autologous pericardial strip[63] is always performed.

After MV repair, the diastolic mitral inflow is assessed by continuous flow Doppler echocardiography. A mean of 2.0-4.0 mm Hg is deemed satisfactory. In all the repair strategies we employed, the minimal final MV opening area should not be less than 10% below norm according to BSA in children. We are cautious to avoid the occurrence of systolic anterior motion (SAM) phenomenon.[63]

2.8.1.6 Mitral valve insufficiency in hypertrophic obstructive cardiomypathy

Delmo Walter EM, Siniawski H, Hetzer R. Sustained improvement after combined anterior mitral valve leaflet retention plasty and septal myectomy in preventing systolic anterior motion in hypertrophic obstructive cardiomyopathy in children. Eur J Cardiothorac Surg. 2009 Sep;36(3):546-552.

Our technique of mitral valve repair in this setting has been modified either to avoid the potential development of SAM after myectomy or treat the existing SAM with MV leaflet repair. We addressed the prolapsing anterior leaflet by avoiding the use of an annuloplasty ring, and performing anterior leaflet retention plasty (ALRP) combined with septal myectomy, which we found to be excellent in restricting MV motion allowing more complete relief of subaortic obstruction and MI, and avoiding SAM.[74,75] Our approach is transaortic septal myectomy to address the left ventricular outflow tract obstruction (LVOTO), with concomitant ALRP performed through a left atriotomy.

Subaortic septal myectomy*:* Intraoperative TEE is performed after induction of general anesthesia, with particular attention to the cardiac anatomy, MV morphology and function, and thickness of the ventricular septum. Exposure is gained through a median sternotomy and direct intracardiac pressures are measured simultaneously in the left ventricle and aorta. If the left ventricular outflow tract (LVOT) gradient is low (<30 mm Hg) because of anesthesia, isoproterenol is administered or premature ventricular contractions are induced to determine the maximal gradient. Standard cardiopulmonary bypass with moderate hypothermia (28-32°C) is used and the left heart is vented. During aortic occlusion, myocardial protection, especially important because of the severe ventricular hypertrophy is given through a generous infusion of antegrade cold crystalloid cardioplegia, followed by additional doses administered selectively onto the right and left coronary ostia every 15-20 minutes. An oblique aortotomy is carried rightward down to the non-coronary sinus and towards the aortic annulus. The aortic valve is inspected and the subvalvular region is exposed. Optimum visualization of the ventricular septum is facilitated by posterior displacement of the left ventricle. Aside from using Morrow technique, which used two parallel longitudinal incisions in the septum, we make parallel incisions into the septum directly opposite the anterior MV leaflet. The resection of long blocks of septal myocardium from between the two incisions is started just below the aortic annulus of the right coronary sinus and the commissure between the right and the left coronary sinuses.[74] Importantly, the incision is continued apically beyond the point of mitral septal contact, marked by a fibrous band. This wide incision beneath the valve improves exposure of the important area toward the apex. In our 25 years' experience, intraoperative pre-septal myectomy pressure gradient to warrant surgical intervention ranged from 40-105 mm Hg (mean 60 ± 25 mmHg), while post-septal myectomy gradient ranged from 0-18 mmHg (mean 5 ±6 mm Hg) when using this technique.

Anterior Leaflet Retention Plasty. Access to the mitral valve is described in sections 3.4, 3.5 and 3.6 After carefully assessing the morphology and mobility of the anterior MV leaflets and the subvalvular apparatus, the segments of anterior leaflet closest to the trigones are sutured to the corresponding posterior annulus wherein the polypropylene mattress sutures are pledgeted with untreated autologous pericardium. These sutures are passed through the coaptation line of the anterior leaflet and the corresponding posterior annulus trigone [74,75] Thus, the mobility of the anterior MV leaflet is limited in its trigonum-near segment, thereby unable to produce systolic anterior motion and mitral insufficiency [74,75]

.

This repair is routinely guided by intraoperative transesophageal echocardiography with focus on the septal anatomy and thickness, MV function, mobility as well as anatomy of the subvalvular apparatus.

After the septal myectomy and ALRP, the aortic and mitral valves are inspected to ensure that they have not been injured. After weaning the patient from cardiopulmonary bypass, pressures are remeasured in the left ventricle and aorta and TEE evaluation is repeated, with special attention to the width of the interventricular septum, the residual LVOT gradient, MI and SAM. If myectomy is successful, there will be little or no residual gradient, and little or no SAM of the mitral valve. In general, we would resume cardiopulmonary bypass for re-resection, if the gradient is greater than 15-20 mmHg. Post-myectomy mitral insuficiency should then be reduced to a regurgitant fraction of 0 to 10%.

2.8.2 Congenital mitral stenosis

2.8.2.1 Shone's anomaly

Understanding the morphology of the mitral valve in Shone anomaly (Fig.7) is critical to determine the reconstructive surgical approach. Parachute mitral valve and mitral ring are the most prevalent variants of mitral stenosis in this disease.[16,18,19,20,76] The fibrous mitral ring, a characteristic feature, rarely occurs as an isolated lesion and is not usually severely obstructive. Resection of this ring should be straightforward. Sharp dissection of the fibrous mitral ring is required to initiate the resection. It is very important to remove all components of the ring. Since the ring is usually within the mobile portion of the leaflet, precautions are taken to avoid injuring the leaflet body itself when dissecting the ring off.

Indications for MV repair are abnormal MV with <2 cm^2 orifice area, and mean resting end-diastolic gradient of >5mmHg, presence of mitral ring and associated LVOT obstructive lesions. Hemodynamic criteria include pulmonary artery pressure of >25 mmHg at rest and >30 mmHg on exertion. Increased left atrial to left ventricular pressure gradient is an indication for repeat MV surgery.

Our approach to the left ventricular inflow and outflow tract obstructive lesions is *single-stage* surgery whenever possible. Single-stage repair is done when all obstructive lesions and intracardiac defects which are deemed significant, are diagnosed all at the same time. This, however, does not preclude eventual reinterventions when necessary. *Staged repair* is done on patients, on whom the usual first operations addressed the left ventricular outflow tract, and the next interventions are done as the other concomitant lesions appeared hemodynamically significant to warrant surgery.[77]

In general, we treat first the most distal obstructive lesions, which is usually coarctation of the aorta.

During MV rsurgery, the mitral annulus, leaflets, chordae tendinae, and papillary muscles are meticulously inspected to determine the precise nature of the stenotic lesion. Anterior and posterior leaflet coaptation is assessed with regards to the presence of sufficient tissues along the coaptation plane The valve orifice area is assessed with a Hegar dilator or with a Ziemer-Hetzer valve sizer, as described in section 3.7. Various repair techniques are employed in accordance to the cause of mitral stenosis and the presenting valve morphology.

In children with fused commissures, commisurotomy is done on both the anterolateral and posteromedial commissures. Division of chordae tendinae and division and splitting of papillary muscles were performed in patients with short, fused and matted chordae.

Repair of parachute valve is previously described in section **2.8.1.5**

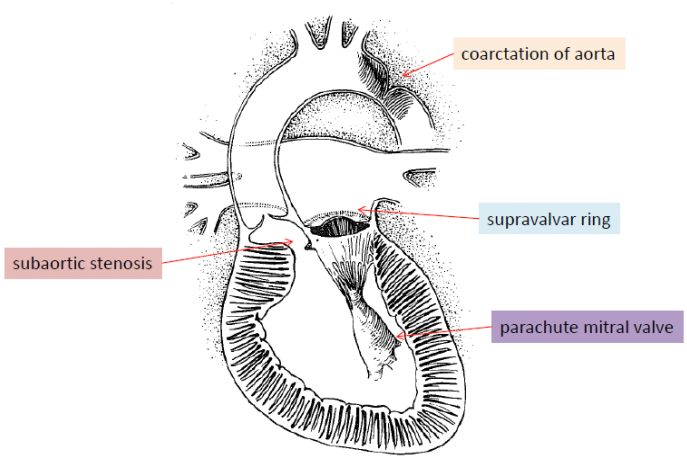


Fig. 7. Graphical illustration of the four obstructive lesions in Shone's anomaly

2.8.3 Acquired mitral valve disease

2.8.3.1 Infective endocarditis

The goals of surgery are to eradicate the focus of infection, to repair destroyed valve structures, to prevent the development of complications, and to prevent relapse of the infection.[78] MV repair has the advantage of preserving the subvalvular apparatus and ventricular function. As ventricular function is impaired at the time of operation, any preservation of function should achieve a reduction of operative mortality, improve the postoperative valve remodeling and increase the long-term survival. All operations are performed as previously described in section 3.4, 3.5 and 3.6. The MV complex are exposed and meticulously inspected to determine the extent of valvular and subvalvular damage from infective endocarditis and to plan the procedure. Our protocol prescribes the preservation of the native valve and avoidance of prosthetic materials whenever possible. Standard

operative principles are adequate debridement of all infected tissues, meticulous washing of all affected areas with 1.5% Povidone containing 10% available Iodine solution irrespective of presence or absence of purulence or vegetations, meticulous removal of vegetations when present, and reconstruction using untreated autologous pericardial strips and pledgets for suture reinforcement.[63,78]

MV repair is performed according to the presenting lesions and consist of any of the following: anterior commissuroplasty,[62,78] posterior commissuroplasty with leaflet resection,[78] Hetzer-modified Paneth technique,[62,78] and chordal rupture is repaired by chordal reimplantation.

We used untreated autologous pericardial pledgets instead of felt material for suture reinforcement. Leaflet perforations, when found, are closed either directly or with an untreated autologous pericardial patch. Meticulous removal of vegetations is done when indicated.

2.9 Mitral valve repair in lesion associated with a primary congenital heart defect (atrioventricular septal defect and a small left ventricle)

Delmo Walter EM, Ewert P, Hetzer R, Huebler M, Alexi-Meskishvili, Lange P, Berger F. Biventricular repair in children with complete atrioventricular septal defect and a small left ventricle. Eur J Cardiothorac Surg 2008;33:40-47.

A pericardial patch is harvested at the beginning of the procedure for closure of the atrial septal defect (ASD). Surgery is conducted through a right atriotomy parallel to the right AV groove, extending from the right auricle to the level of the entrance of the inferior caval vein. The remains of the atrial septum is incised up to the atrial wall to avoid tension and deformation of the left-sided (mitral) valve which would later provide optimal suspension and mobility of the reconstructed valves.[79] Cold saline solution is used to fill the ventricular chambers and float the AV valves into a closed position to evaluate valve anatomy, establish the line of coaptation between the superior and inferior components of the valve and identify the proper line of division into right and left parts of these components.[79] The AV valves consisted of basically five-leaflet common atrioventricular valve, with a common orifice. A stay suture is placed to mark the point that effectively joined the antero-superior and postero-inferior bridging leaflets.[79]

In this special condition of a small left ventricle, we do a biventricular repair[79] consists of a double-patch technique. To enlarge the orifice of the MV, the ventricular septal patch is placed more to the right of the ventricular crest. Since the ventricular component is small, the ventricular septal

defect (VSD) is closed by sewing multiple interrupted pledgeted 5-0 Tevdek sutures more onto the right side of the defect and individually sutured to the inferior portion of a precisely- fashioned semi-oval Weavenit patch and then tied.[79] The aforementioned patch is then inserted cautiously to avoid chordal distortion. Care is taken to avoid potential injury to the conduction tissues. Another set of interrupted sutures are then passed through left superior and inferior common bridging leaflets to partition the valves into right and left components at the points where the leaflets naturally abutted the crest of the interventricular septum, and then through the leading edge of the VSD patch bringing the antero-superior and postero-inferior leaflet components closer together and increase the area of central coaptation.[79] These sutures are then individually passed through the previously harvested and untreated autologous pericardium, which will later be used to close the ASD. They are then firmly tied, which now obliterates the ventricular component of the defect by approximating leaflet tissue to the septal crest.[79] The distance from the left-sided AV (mitral) valve level to the crest of the interventricular septum is accurately assessed to reconstruct the left-sided AV (mitral) valve at the appropriate height to avoid LVOTO. Both AV valves are now sandwiched between ventricular and atrial patches with interrupted monofilament sutures. The stay suture joining the left superior and inferior bridging leaflets are then used to initially close the cleft in the anterior leaflet of the mitral valve. Another stay suture is placed at the point of the first papillary muscle-derived chordal support of the cleft and placed on tension to facilitate apposition of the two leaflet components and avoid purse-string-like effect.[79] The cleft between the left-sided superior and inferior bridging leaflets is closed completely and may be partially closed if there is a tissue deficiency and left-sided obstruction, i.e membrane or muscular hypertrophy. Thus, when necessary, simultaneous membranectomy and myectomy of the LVOT is performed. Closure of the cleft is done in an interrupted fashion with Prolene 7-and reinforced with pericardial pledgets, when the leaflet tissue appears friable. During cleft closure, a minimal acceptable mitral valve diameter is maintained according to patient age to avoid mitral stenosis. To assess the quality of the valve repair, cold saline solution is injected into the left ventricle, and if necessary additional interrupted sutures is placed on the cleft.[79] Attempts to gain leaflet tissue area for the mitral component, to bring the left superior and inferior leaflet component closer together and to increase the area of central coaptation, are done by extending the ASD further towards the atrial wall, enabling the left-sided AV valve leaflets to fall downward, avoiding restriction of anterior leaflet mobility as well as increasing the valve surface area and preventing leaflet puckering, thus improving leaflet coaptation. The ASD is then closed with the previously harvested untreated autologous pericardial patch with a running 6-0 Prolene suture starting in the commissure between the right mural leaflet and the inferior bridging leaflets, hence, avoiding placement sutures along the crura on both sides. Suturing continues along the free edge of the right atrial wall to the bottom of the coronary sinus in order to avoid the AV nodal area and preserve the coronary sinus drainage to the right atrium.[79] From hereon, the border of the ASD is followed to reach the superior

AV ring again. The right AV (tricuspid) valve is floated into a closed position, and, if indicated, the right side coaptation area of the superior and inferior bridging leaflets is closed with interrupted Prolene 6-0 sutures.[79] Valve competence is repeatedly tested with saline injection through the valve orifice.[79]

3.0 Original Studies

3.1 Mitral valve repair techniques and long-term outcome of repair in congenital mitral valve insufficiency in infants and children

1. **Delmo Walter EM,** Komoda T, Siniawski H, Hetzer R. Surgical reconstruction techniques for mitral valve insufficiency from lesions with restricted leaflet motion in infants and children. Journal of Thoracic and Cardiovascular Surgery 2012,143:S48-S53.

*This paper was presented at the American Association of Thoracic SurgeonsS 2011 Mitral Conclave: New York , USA, May 5-6, 2011

Impact Factor 3.610

2. Hetzer R and **Delmo Walter EM**. Repair of congenital mitral valve insufficiency In Bradley S (ed). Operative Techniques in Thoracic and Cardiovascular Surgery Winter 2010:15(4);260-272.

Because congenital mitral valve insufficiency (MI) from restricted leaflet motion is clinically important although a rare entity, various repair techniques and results have been reported.[68-73] Limited durability of repair has been a major setback. With meticulous intraoperative assessment of valve morphology and appropriate selection of surgical strategy, we believe repair can be long-lasting. In our institution, MV repair is the preferred technique for any kind of mitral disease in childhood. Repair allows for valve growth along the anterior leaflet annulus,[80,81] provides no anticoagulation and offers no thrombotic risk. This is best achieved by a spectrum of repair techniques tailored to the presenting lesion. We do not employ any prosthetic material and we stabilize our repair with untreated autologous pericardial strip.

In the first paper, we reported our 23-year experience with leaflet and annular remodeling techniques in 49 infants and children with congenital MI from lesions with restricted leaflet motion.

These 49 children, classified according to age groups , underwent MV repair for severe MI from restricted leaflet motion (Type III Carpentier's functional classification). In the two reoperations within a year after the initial repair. it was remarkably noted that the mitral annulus was markedly

dilated, the anterior mitral leaflet as was noted to be slightly thickened, pliable and freely mobile but the commissural portion of the posterior annulus was noted to be enlarged in one patient, and the leaflets were severely retracted without any coaptating potential, in the other; however, the pericardial strip with the plication sutures was perfectly attached to the posterior annulus and covered by a layer of fibrous tissue without calcification and each pericardial strip appeared completely endothelialized and was indistinguishable from the atrial endocardium.

Mean duration of follow-up was **11.2±7.2** (median 11.28, range 6 months-23.7) years. Postoperatively and on serial follow-up, all patients had a tremendous abatement of MI with a mean mitral inflow gradient of 2.81±0.58 mm Hg (acceptable maximal gradient in children is 3.5 mm Hg) and all have functionally improved from NYHA Class III to Class I .

Age has not been found to be a significant risk factor for reoperation and mortality nor the surgical repair techniques employed.

The surgical outcome is satisfactory. Freedom from reoperation was 100% at 30 days and 95.7±3.0%, 86.0±5.3%, 79.6±6.6% and 71.1±8.1% at 1, 5, 10, 15 and 20 years, respectively (Fig. 8). Stratified based on age groups at the time of initial repair, freedom from reoperation was 100% at 30 days in all age groups (Fig. 9) . In ***<1 year old*** , freedom from reoperation was 100% at 1 year and 57.1±8.7% at 5 and 20 years; it was 90.9±6.1% at 1 and 5 years, 78.8±9.6% at 10 years and 63.2±12.6% at 15 years in age group ***>1-5 years old***; 100% at 1 year and 90.9±8.7% at 5, 10 15 and 20 years in those belonging to age group 6-12 years old. There has been no reoperations performed in those children >13 years old at the time of initial repair.

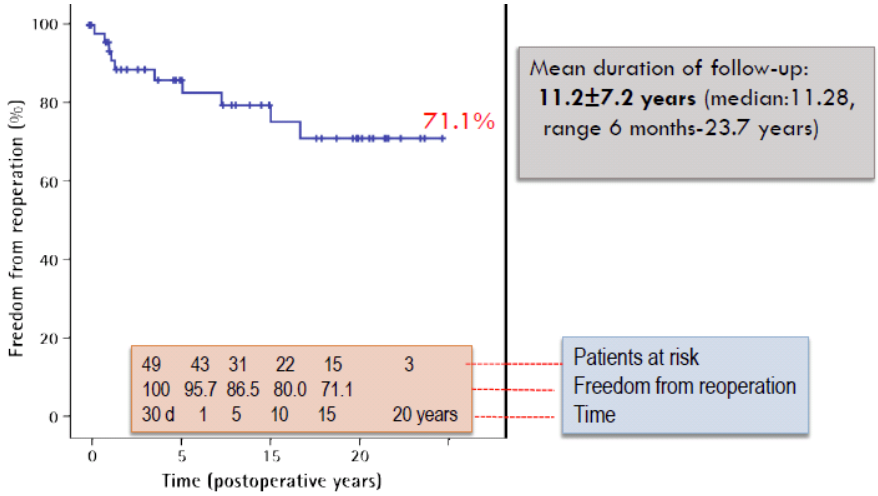
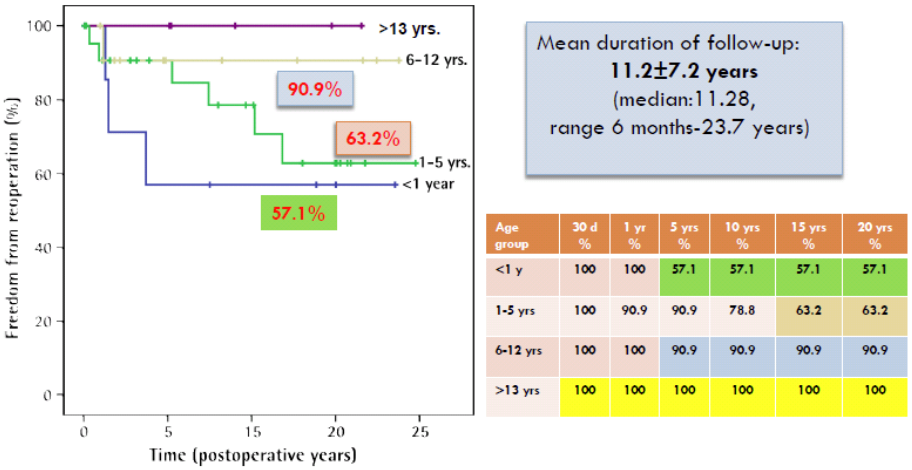


Fig. 8. Overall freedom from reoperation after mitral valve repair for MV insufficiency from lesions with restricted leaflet motion.



Age group	30 d %	1 yr %	5 yrs %	10 yrs %	15 yrs %	20 yrs %
<1 y	100	100	57.1	57.1	57.1	57.1
1-5 yrs	100	90.9	90.9	78.8	63.2	63.2
6-12 yrs	100	100	90.9	90.9	90.9	90.9
>13 yrs	100	100	100	100	100	100

Fig. 9. Freedom from reoperation after mitral valve repair for MV insufficiency from leasions restricted leaflet motion, classified according to age groups.

Cumulative survival was 100% at 30 days, 1 year and 5 years and 96.9±3.6% at 10,15 and 20 years (Fig. 10).

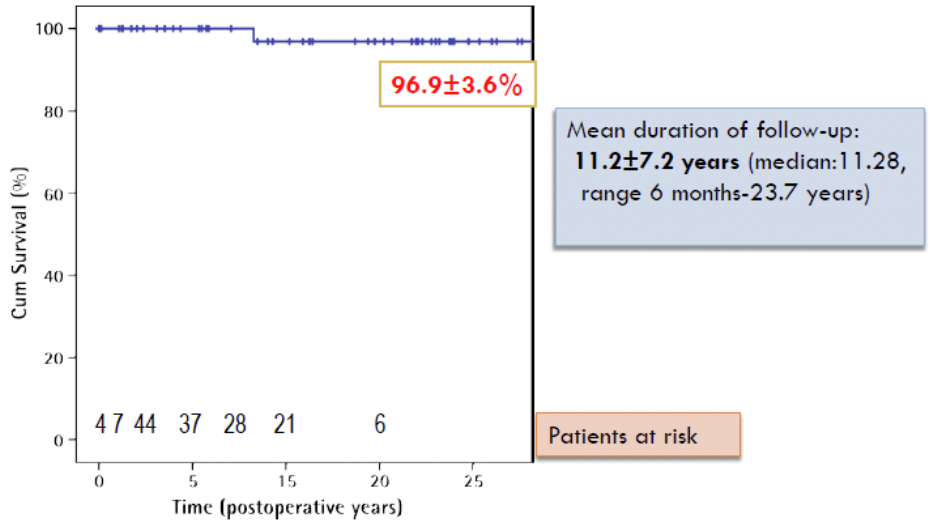


Fig. 10. Cumulative survival of infants and children who underwent mitral valve repair for mitral valve insufficiency from restricted leaflet motion

The ***principles of mitral valve repair*** for congenital mitral valve insufficiency in childhood is reported in the second paper. We emphasized that the concept of repair is aimed primarily at growth of the patient to an age when an adult-sized prosthesis can be implanted, if necessary. We highlighted that the primary goal in repair is to achieve complete and rapid closure of the mitral orifice by a well-mobile naterior leaflet and a sufficiently large coaptation area by bringing the posterior leaflet closer to the anterior leaflet. This is accomplished by repair techniques aimed at functional rather than

anatomical restoration of the incompetent valve. We also accentuated the use of autologous pericardial strip to stabilize our repair. Not only various repair techniques, which were either modified from established existing valve repair techniques or developed in our insitution, but also several important points are featured in this paper, like approach to the mitral valve, its exposure and evaluation of adequacy of repair. Precautions to prevent systolic anterior motion has been a significant feature in this paper.

3.2. Mitral valve repair techniques and long-term outcome in mitral valve insufficiency in hypertrophic obstructive cardiomypathy

Delmo Walter EM, Siniawski H, Hetzer R. Sustained improvement after combined anterior mitral valve leaflet retention plasty and septal myectomy in preventing systolic anterior motion in hypertrophic obstructive cardiomyopathy in children. Eur J Cardiothorac Surg. 2009 Sep;36(3):546-552.

*This paper was presented at the 22nd EACTS Meeting, September 13-18, 2008, Lisbon, Portugal

Impac tFactor 2.397

There has been few reports in surgical management of mitral valve in children with HOCM.[82] Mitral valve reconstruction techniques complementing septal myectomy to prevent postmitral repair SAM such as triangular resection of the anterior leaflet,[83] sliding plasty of the posterior leaflet,[84] edge-to-edge-technique,[85] anterior mitral leaflet extension with a pericardial patch,[86,87] radical debridement and repositioning of the papillary muscles [88] as well as complete excision of secondary chordae of the anterior mitral leaflet[84] may be useful. MV replacement is not desirable in children, because of its attendant morbidities as well as lack of suitable prosthesis appropriate for age.[63] We perform anterior mitral leaflet retention plasty (ALRP) [74.75] in combination with septal myectomy in children with HOCM and we investigate the long term results of these combined procedures in 12 children.[74]

Most patients in this group had moderate to severe mitral regurgitation (2.82±0.18). All patients had typical marked SAM of the anterior mitral valve leaflet (mean grade 3.08±0.61). Only 1 (8.3%) had an associated ventricular septal defect, in whom direct closure was performed during the septal myectomy and ALRP procedure.[74]

With the repair techniques employed, there was no reoperation for repeat myectomy nor repeat mitral valve repair or replacement. There was no early nor late mortality. At a mean follow-up period was 13.18±1.22 years (range 9 months to 15 years), survival is 100%. During the follow-up period, patients were either asymptomatic (33.3%) or had mild symptoms (50% and 16% in NYHA functional Class I and II, respectively). The number of cardiac drugs prescribed decreased significantly. Moreover, the LVOT gradient as well as MI was significantly reduced after surgery and further reduced during the succeeding follow-up period. The most significant finding is that there is no SAM immediate postoperative and during the latest follow-up. At the latest patients' follow-up, these results were sustained. [74]

As there was no reoperation and mortality, no risk factors have been analyzed.

3.3 Surgical repair techniques and operative outcome in congenital mitral valve stenosis

Delmo Walter EM, Komoda T, Siniawski H, Miera, O, Van Praagh R, Hetzer R.. Long-term surgical outcome of mitral valve repair in infants and children with Shone's anomaly. European Journal of Cardiothoracic Surgery 2012. *Article in Press*

* This paper was presented at the 25th EACTS Annual Meeting, October 1-5, 2011, Lisbon, Portugal

Impact Factor 2.397

The finding in Shone's [10] original description that the extent of mitral valve (MV) involvement seems to be the predominant factor determining outcome is supported by the review of 30 cases by Bolling and colleagues [18] which represents the most comprehensive report to this date, and comprised patients with a multitude of anatomic variants and different management approaches. That outcome is related to the predominance and severity of the mitral component of the disease has also been the finding in the report by Brauner et al [19] Brown et al [20], and Ikemba et al [76] who published mitral valve morphology and morbidity/mortality in 50 patients with Shone's anomaly.

Having collected a large series of Shone's anomaly with the longest follow-up, we analyzed the operative results and long-term outcome of mitral valve repair techniques performed to correct left ventricular inflow tract lesions of this-congenital anomaly.[77]

There were 45 children with Shone's anomaly who underwent surgery at the Deutsches Herzzentrum Berlin.[77]

These children were divided into age groups: **<1 year**, n=12, mean age 0.4±0.27 (median 0.32, range 0.02-0.96) months; **1-5 years**, n =19, mean age 3.5±1.4 (median 3.6, range 1.1-4.8) years; ***>5-10* years,** n=8, mean age 8.6±2.0 (median 8.6, range 6.2-9.2) years; and ***>10 years***, n=6, mean 13.2±1.6 (median 15.2, range 11.2-16.8) years. The majority of patients in this series presented initially in the neonatal period with coarctation of aorta (n=12) and in the 1-5 year old group (n=19) as the predominant outflow obstructive lesions. The neonatal coarctation presented with severe symptoms which could have masked the other intracardiac pathology until it was repaired. All 45 children, on whom MV surgery was performed at any stage in their disease, had significant transmitral gradient justifying the necessity of intervention. Since there were no patients with hypoplastic, small or even borderline left ventricular, all patients had biventricular repair.

Mean duration of follow-up was 17.5±1.5 (median 11.3, range 6.4-22.7) years.

Freedom from reoperation was 97.6±2.4%, 89.3±5.1%, 77.1±7.2%, 72.0±8.3% and 52.8±11.8%, at 30 days, 1, 5, 10 and 15 years postoperatively, respectively.[77]

Stratified based on age groups at the time of initial repair, freedom from reoperation was 100% at 30 days in all age groups. In the ***<1 year olds***, freedom from reoperation was 95.56 % at 1 year and was sustained until the late follow-up period. Repeat MV repair was performed mostly in the 1-5 year age group until 10 year follow-up. In the ≥10 year-old group it was noted that there was no repeat MV surgery 5 years after the initial MV repair.[77] We performed only one MV replacement, and this was on a patient with parachute valve (see section on late mortality).

Multivariate analysis of perioperative risk factors for reoperation and mortality revealed that the types of MS [type I congenital MS with fused commissures and thickened leaflet (p=0.004), hypoplastic MV (Type II), with emphasized shortened chordae and miniature papillary muscles (p=0.001), supravalvular mitral ring (p=0.002) and parachute valve (p=0.003)], the associated pulmonary hypertension (p=0.001) and the left ventricular outflow tract lesions (p=0.00) are significant risk factors. In this paper, we emphasized that the repair techniques as well as staged surgery, however, proved to be non-influential to the outcome in this study population.[77]

3.4 Surgical techniques and operative outcome of repair of mitral valve in infective endocarditis

Delmo Walter EM, Musci M, Nagdyman N, Hübler M, Berger F, Hetzer R. Mitral valve repair for infective endocarditis in children. Ann Thorac Surg. 2007;84(6):2059-65.

Impact Factor: 3.039

Children with untreated congenital cardiac disease (CHD), especially those with ventricular septal defect and patent ductus arteriosus, as well as children with complex intracardiac abnormalities undergoing surgical palliation or definitive correction [89] are considered to be at higher risk [90,91] for developing infective endocarditis. Although improved outcomes have been reported over the past decades, infective endocarditis in infancy and childhood remains a most serious condition. At Deutsches Herzzentrum Berlin, we have seen only 8 cases of IE in children during the last 25 years.

This paper described our own experience with mitral valve endocarditis in children in whom MV repair was performed.[78]

In this small series, we have shown that mitral valve repair in children with infective endocarditis is safe, with no operative and long-term mortality. All patients had good functional status during long-term follow-up and not one had undergone mitral valve reoperation.

Emphasis was made on the goals of surgery which are to eradicate the focus of infection, to repair destroyed valve structures, to prevent the development of complications, and to prevent relapse of the infection. Mitral valve repair also has the advantage of preserving the subvalvular apparatus and ventricular function. As ventricular function is impaired at the time of operation, any preservation of function should achieve a reduction of operative mortality and improve postoperative remodeling and long-term survival. There was no morbidity. Actuarial freedom from reoperation and actuarial survival rate at 1, 5, 10 and 15 years were 100%. [78]

3.5 Surgical techniques and operative outcome of mitral valve repair in lesion associated with a primary congenital heart defect (atrioventricular septal defect and a small left ventricle)

Delmo Walter EM, Ewert P, Hetzer R, Huebler M, Alexi-Meskishvili, Lange P, Berger F. Biventricular repair in children with complete atrioventricular septal defect and a small left ventricle. Eur J Cardiothorac Surg 2008;33:40-47.

Impact Factor: 2.181

In complete AV septal defects, the left atrioventricular valve, which is the mitral valve, has a cleft, and may be dysplastic, with stiffened, thickened valve tissues, curling-in of the leaflets, shortened chordae and annular dilatation.[92] The valves can also be severely deformed with fenestration and thickenings.[93] Mitral valve insufficiency is moderate to severe, justifying the need for early correction. Performing a mitral valve repair and closure of the cleft in the context of a small ventricle, could pose a problem. It is not only an issue of whether to partially or fully close the cleft, so as not to produce iatrogenic mitral stenosis, but also we must be aware that repairing the MV structural abnormalities may produce obstruction in the left ventricular outflow tract. In the background of a small left ventricle, this is a challenge. Thus, it is also much an issue of whether to perform single-ventricle palliation or biventricular repair. Hence, surgical decision-making depends not only on the left-sided (mitral) valve but also on the size of the left ventricle.

In this paper, we investigated and analyzed the outcome of mitral valve repair in the presence of a small ventricle in 19 patients with complete atrioventricular septal defect who underwent biventricular repair.[79]

A small left ventricle is defined on the basis of angiographic measurements based on a model developed at Deutsches Herzzentrum Berlin, expressed as the left ventricle/right ventricle long axis ratio <1.1[79] where the length of the left/right ventricle is measured in the long axis during ventriculography. [79]

The left-sided AV valve morphologic features were specifically noted, particularly the shape of the leaflets, the arrangement of the subvalvular apparatus at the ventricular surface of the leaflet, and the pattern of division of the tendinous cords as they arise from the papillary muscles in the operative records of these 19 patients.

We lost two patients (10.5%, LAR of 0.45 and 0.60) on the 8th and 11th postoperative day, respectively, from intractable pulmonary hypertension. The third patient with an LAR of 0.62 had ECMO support for postoperative myocardial failure and subsequently underwent successful heart transplantation on the 21st postoperative day. This patient is doing well 7 years after transplantation.

Univariate analysis of gender, age at operation, weight, Down's syndrome, LV/RV LAR; prolonged postoperative ventilation, and postoperative left side AV valve regurgitation was performed. It revealed ***postoperative left-sided valve regurgitation*** as the only independent negative risk factor for reoperation.[79] Univariate analysis of mortality showed LAR <0.65, pulmonary hypertensive crisis and ECMO support to be associated with death (Table 2, Original studies, p.81). The Mann-Whitney U-test showed no statistical differences in bypass or cross-clamp times with regards to mortality and reoperation.[79]

Follow-up was complete and comprised 1114 month patient-years (mean 58.6±9.12 months, range: 0-150 months. The postoperative course of the 17 surviving patients was uncomplicated and they are seen regularly and transthoracic echocardiography is performed to assess ventricular function, valve function, and the status of the left ventricular outflow tract. There has never been any reoperation for residual VSD, residual ASD, subaortic stenosis or mitral valve dysfunction. The long-term survivors (89.5%) with LAR >0.65 (0.80± 0.11) undergo regular follow-up with good left ventricular function as assessed by echocardiography. At the latest follow-up, mitral regurgitation was non-existent in 13 (81.25%) and mild in 3 (18.75 %) patients.

3.6 Modified surgical techniques and long-term outcome of mitral valve repair in childhood

Hetzer R, **Delmo Walter** EM, Hübler M, Alexi-Meskishvili V, Weng Y, Nagdyman N, Berger F. Modified surgical techniques and long-term outcome of mitral valve reconstruction in 111 children. Annals of Thoracic Surgery 2008;86(2):604-13.

Presented at the 43rd Annual Meeting of the Society of Thoracic Surgeons, January 27-31, 2007, San Diego, California, USA.

Impact Factor 2.689

In this paper, we reviewed our 19-year experience with MV repair in pediatric age group to assess the effectiveness and outcome of our repair techniques and determine early and long-term survival and freedom from reoperation and valve replacement. Described in detail are the various repair techniques applied according to presenting mitral valve morphology of 111 children with mitral valve lesions classified according to Carpentier's functional classification. [63]

Multivariate analysis revealed that age at operation, concomitant operations, urgency of operation, operative times and valve status on discharge were risk factors for mortality and reoperation.[63]

Overall freedom from reoperation was 88.8± 3.1, 86.2± 3.5, 79.2± 5.1 and 73.5± 7.2 at 1, 5, 10 and 15 follow-up years.[63]

Freedom from repeat MV reconstruction was 96.3±1.8%, 96.5±2.6%, 91.1±1.5% and also 91.1±1.5% at 1, 5, 10 and 19 years, respectively.[63]

Overall freedom from MV replacement is 96.3±1.8%, 93.2±2.8%, 87.4±4.9% and 81.1±7.5% at 1, 5, 10, and both 15 and 19 years, respectively.[63]

Actuarial survival was 95.5±2.6% at 30 days, 88.4±3.2% at 1 year, 85.5±3.7% at 5 years and 77.4±5.1% at both 10 and 19 years.[63]

Three patients from foreign countries were lost to follow-up at 4, 9 and 13 years. Follow-up of the remaining patients was complete and comprised 593 patient-years (mean 5.4±0.46 years). Improvements in degree of both insufficiency and stenosis after valve reconstruction were maintained until the last follow-up. An acceptable postoperative outcome was achieved in all 85 (76.6%) surviving patients. Mild stenosis was noted in 3 of the 13 patients who previously had severe MS. Among the 72 patients who had had MI as the predominant lesion, only 8 patients had mild to moderate insufficiency at their latest follow up.[63] The surgical techniques employed for each presenting MV lesion have been proven to be long-lasting.

3.7 Mitral valve growth after repair of mitral valve insufficiency

Delmo Walter EM, Siniawski H, Ovroutski S, Hetzer R. Mitral valve growth after posterior annular stabilization with untreated autologous pericardial strip in children with mitral valve insufficiency. Ann Thorac Surg. 2010 Nov;90(5):1577-85; discussion 1585.

*This paper was presented at the 46th Annual Meeting of the Society of Thoracic Surgeons, January 25-27, 2010, Fort Lauderdale, Florida, USA.

Impact Factor: 3.644

Because repair of MV lesions in infants and children are preferred, regardless of the etiology and morphology of MV lesions, we became interested in whether the repair techniques we applied allows the growth of the mitral valve as the child grows. The technique we adopted involves the use of untreated autologous pericardium for posterior mitral valve annuloplasty to achieve an annular reduction with a flexible tissue, to ensure leaflet coaptation and to support the valvuloplasty. Several studies have shown the significance of preserving the anatomic characteristics of the mitral annulus, which provides an active contribution to the physiology of the valvular apparatus throughout the cardiac cycle.[27,28,94,95] A complete understanding of the function of the mitral annulus is extremely essential in deciding what type of repair technique would be valid. In our instititution, we are convinced that leaving the autologous pericardium untreated makes it more flexible to maintain the physiologic movement of the mitral annulus, construct a soft reinforcement which conforms to the natural tri-dimensional geometry of the mitral annulus, offers long term durability, and absence of thrombogenicity and calcification, as evidenced during reoperations. Stabilization of the posterior annulus with untreated autologous pericardium has positive influence on left ventricular function while preserving the contractile properties of the mitral annulus. These benefits in addition to the easily accomplished surgical technique, the effective functioning of the remodelled valve, the preservation of the natural shape of the valve and allowing it to grow as the patient grows older, make this technique a useful surgical tool for mitral valve reconstructive surgeries.[81]

In this paper**,** we aimed to evaluate whether our technique of posterior annulus stabilization with untreated autologous pericardial strip allows growth and remodelling of anterior MV leaflet and its annulus, in infants and children with congenital mitral insufficiency.

We prospectively followed-up seventeen children after MV repair for mitral insufficiency from severe annular dilatation, anterior mitral leaflet prolapse and chordal rupture, and parachute mitral valve.[81] MV repair techniques were Paneth-Hetzer posterior annuloplasty [81] and Gerbode-Hetzer plication plasty,[81] both modified with pericardial strip stabilization of the posterior annulus. Echocardiograms were taken perioperatively and serially during follow-up.

During the mean follow-up period of 13.62±2.37 years, the mitral valve area, the annular size and the anterior leaflet area, have increased significantly within the *z* score. The anteroposterior diameter of the annulus contour exhibited a slight thickening, which implies that the pericardial strip stabilized the posterior annulus while allowing growth, without inducing mitral insufficiency[81].

Indeed, we were able to prove that stabilization of posterior shortening annuloplasty *allowed growth of the leaflets and annulus which is commensurate to the somatic growth* of the patient. Measurements of the ventricular diameter showed a significant increase during systole. The substantial increase in mitral valve area appropriate for the linear gowth of the patient implied mitral valve growth. The mean annular size increased and planimetry revealed a mild but significant increase in the growth of the annulus. The anteroposterior diameter of the annulus contour exhibited a slight thickening which implies that the pericardial strip stabilized the posterior annulus while allowing growth, without inducing mitral insufficiency. Likewise, a highly significant improvement in left ventricular function was also evident.

In the three reoperations, intraoperative finding included a markedly dilated annulus, with dysplastic leaflets but without any structural defects of the subvalvar apparatus. Remarkably, the pericardial strip was perfectly attached to the posterior annulus, covered by a layer of fibrous tissue without any calcification, intact, completely endothelialized and was indistinguishable from the atrial endocardium.

4.0 Discussion

A precise and detailed understanding of the surgically relevant anatomy and morphology of the mitral valve complex, a thorough knowledge of the pathophysiology of each mitral valve lesion, and establishing principles of mitral valve repair in infants and children, are undoubtedly the basic prerequisites in conceiving and modifying surgical techniques aimed to restore its functional competency with a good long-term outcome, while allowing growth of the valve. It is exceptionally arduous to repair the mitral valves in infants and children primarily because the leaflets and subvalvar apparatus are small, immature and fragile. However, we believe that the only best option in this population is to repair the mitral valve.

4.1 Surgical techniques and long-term outcome of repair incongenital mitral valve insufficiency in infants and children

In congenital mitral valve insufficiency from restricted leaflet motion, one may be tempted to resect the tissues, and reconstruct it with prosthetic rings, or simply replace the valve, to restore the mitral competence. Various techniques from various authors and institutions have evolved to correct the problem.[9-14,69,96-101] In this population, however, ring annuloplasty would disturb the growth of the mitral valve along its anterior annulus. Valve replacement poses clinical and technical difficulties because of the small-sized annulus, left atrium and left ventricle along with accelerated tissue calcification and degeneration of the bioprosthesis, lifetime anticoagulation, notwithstanding the fact that there is no suitably-sized prosthesis in this age group, and re-replacement of the prosthesis with growth of the child is inevitable.

In deciding to repair the MV with restricted leaflet motion, our primary aim is to achieve a complete and rapid closure of the mitral orifice by a well-mobile anterior leaflet with a sufficiently large coaptation area by bringing the posterior leaflet closer to the anterior leaflet. Primary repair must be achieved by sutures only. The pericardial strip does not lead to further annulus shortening; but stabilized the suture-dependent repair and increase the height of the posterior leaflet coaptation surface area. Moreover, we believe that suture repair techniques for congenital MI allows undisturbed somatic and valve growth,[81.82] delays, if not prevent, the need for future valve replacement, and avoids anticoagulation. Modifications in repair techniques and the routine use of intraoperative TEE to assess the adequacy of repair have significantly contributed to the success of MV repair as the surgical management for MI from restricted leaflet motion in children. Our freedom from repeat MV reconstruction and freedom MV replacement at >20 years' follow-up is highly satisfactory.

4.2 Surgical repair techniques and operative outcome of repair in mitral valve insufficiency in hypertrophic obstructive cardiomypathy

In hypertrophic obstructive cardiomyopathy [88,102,103] variable degrees of LVOT obstruction induced by thickening of the interventricular septum as well as due to systolic anterior motion (SAM) of the anterior mitral valve leaflet are present. SAM is responsible for concomitant mitral insufficiency typically directed postero-laterally into the left atrium.[86] HOCM often presents with anatomic changes of the mitral valve such as increased mitral leaflet area, elongation and pronounced laxity, as well as anterior displacement of the papillary muscles.[104-106] These structural abnormalities are not corrected by septal myectomy alone, and thus patients develop propensity to a residual SAM, persistence of outflow obstruction and mitral regurgitation after septal myectomy leading to a suboptimal outcome.[107,108]

In HOCM, the abnormal motion of the anterior MV leaflet in systole plays a key role in creating the outflow obstruction. The pull and push mechanism of the leaflets due to either increased mitral leaflet length and laxity allow the valve to protrude in the outflow tract. Accordingly, inward displacement of the papillary muscles towards each other results in chordal slack in the central leaflet portion and consequently SAM in the central portion of the valve.[105] To counteract these forces, we developed the technique of anterior leaflet retention plasty (ALRP), wherein the segments of anterior mitral leaflet closest to the trigones are sutured to the corresponding posterior annulus, we restrict the buckling part of the anterior leaflet, restoring the normal mobility of the anterior mitral valve leaflet. This decreases the length of the leaflet which may cause a lateral shift of the chordae attaching prolapsed portion of the leaflet, which in turn stretches the chordae, erects them, and enhances leaflet coaptation. Likewise the normal anatomy of the mitral valve is maximally restored. In fact, the retained anterior leaflet appears tightened in systole, is directed toward the left atrium, and SAM and mitral regurgitation are abolished. This approach is designed to avoid mitral valve replacement, and hemodynamic results are encouraging. The selection criteria for an anterior leaflet retention plasty included not only a relatively thick ventricular septum, presence of an abnormal mitral valve apparatus and prolapse of anterior leaflet, but also a persistent obstruction after myectomy. It is very important to evaluate the presence of abnormalities of the subvalvular apparatus in HOCM, since failure to recognize and treat them may be fatal or lead to incomplete relief of obstruction.[109]

In our experience, adequate myectomy maybe achieved in essentially all patients. However, careful hemodynamic assessment after weaning from the cardiopulmonary bypass is obligatory for optimal results. To counteract residual SAM, several authors [109-112] reported MV replacement as alternative procedure in patients regarded as less-suitable candidates for septal myectomy. While we agree that the mitral valvular dysfunction is an integral factor in producing outflow obstruction, replacement of

the valve is not essential either for reduction of the outflow gradient or alleviation of mitral insufficiency.

ALRP in combination with septal myectomy is an effective and safe treatment for children and adolescents with HOCM. Long-term follow-up shows sustained absence of systolic anterior motion, attenuation of mitral regurgitation, sustained improvement in functional status, and absence or reduction of outflow tract obstruction. Because the present technique offers a broadening of the surgical possibilities, we believe that ALRP could become the preferred choice in persistent LVOT obstruction and SAM.

4.3 Surgical techniques and long-term outcome in congenital mitral valve stenosis (Shone's anomaly)

The formidable surgical challenge presented by children with Shone's anomaly is amplified by the coexistence of restrictive and often surgically unfavorable MV morphology with other obstructive lesions at an early age. The optimal surgical approach to patients with Shone's anomaly is governed by the morphology of each obstructive lesion. We favored an aggressive approach to MV pathology, i.e. we performed mitral valve repair on all patients with transmitral gradient >5 mmHg, which in this population, were underscored.

Mitral ring, a characteristic feature, is the most prevalent cause of mitral stenosis in this disease.[41,113] Its recurrence, though rare, has been described.[20,41,114] Our experience in 12 patients who underwent repeat resection of mitral ring may not be absolutely described as recurrence, because during the repeat resection we found that the rings were mostly of membranous type, completely adherent to the MV leaflet, which may have been difficult to dredge up during the first intervention, owing to absence of a clear line of distinction from the leaflet to which the ring was attached, so that safe dissection was doubtful. We might have possibly missed this membranous part of that fibrous ring, having been overly cautious not to damage the leaflet.

The typical congenital mitral stenosis, was ingenuous to repair. Simple commissurotomy, chordal division and splitting the papillary muscles created an effective orifice area providing relief of the stenosis. We did not see the need to touch the thickened and rolled leaflets but left them as they were seen.

Determining the approach to the hypoplastic MV is equally challenging. It was a dilemma to be faced with a minuscule sized-leaflet and subvalvar apparatus in addition to the immensely delicate and underdeveloped, if not insufficient, tissues, particularly in infants. Repair is the only available option.

With meticulous inspection, especially of the subvalvar apparatus, we were convinced that conscientious division of chordae tendinae to provide adequate interchordal spaces and scrupulous fractionation of the papillary muscles to provide a fairly adequate functional size and length, could provide a functional MV, albeit not long-lasting.

Because repair is often temporary and/or ineffective, MV replacement is undertaken as a last option when repair fails.[115] We agreed with several reports [4, 19,20,76,116] including some of ours [63,64,74] which reported that long-term results of valve repair are superior to replacement in patients with congenital MV anomalies. The use of modified surgical repair techniques tailored according to the presenting MV morphology and the routine use of intraoperative echocardiography have facilitated our confident performance of repair. Brauner and colleagues [19] reported that the failed MV repair procedures in their series were performed before the availability of TEE, and hence it is assumed that this outcome is preventable. We are fortunate to have employed this diagnostic modality ever since its introduction.

Since the MV lesion is not isolated, there has not been any concern about the inadequate loading of the left ventricle which create an impedance to left ventricular ejection, decreasing cardiac output, and inability to sustain postoperative hemodynamics, Creation of a widely patent and competent left ventricular inflow, which is the mitral valve, leads to normalization of postoperative pulmonary pressures and adequate ventricular workload. The results of repair approach in this group are encouraging and we believe that this group gains maximally from repair.

The single stage operative approach did not prove to have a significant positive efffect on long-term outcome in these patients, even in terms of reoperation, since relief of mitral stenosis unmasked any existing left ventricular inflow tract lesions.

We do not aim for an anatomical restoration of the mitral valve; rather we aimed for restoration of functional competency of whatever valve apparatus is involved. We are not able to compare the different types of mitral valve repair to find out whether one is better than the other because these techniques were modified accordingly with time, hence, could not be recognized as an independent risk factor. The reoperation rate for every age group was varied, and different factors were involved such as the type of lesions the patient first had. Our results have shown that the type of mitral stenosis comprise mostly the risk factors determining outcome .

Whatever the type of left ventricular inflow tract lesions are and however they are presented, we believe that even in infants and in very young children, an aggressive repair approach to mitral valve surgery is gratifying.

In this series, repeat MV repair was mostly performed in the 1-5 year age group because of residual mitral dysfunction which could have been greatly intensified when the other left ventricular outflow tract obstructive lesions became pronounced after the intial relief of mitral stenosis.

In all these MV lesions in Shone's anomaly, successful repair allowed for valve growth relative to body growth without the need of anticoagulation.[81] It is important to note that all patients were in modified Ross/NYHA functional class I with normal growth and development.

4.4 Surgical techniques and operative outcome of repair of mitral valve in infective endocarditis

Our study definitely demonstrated that mitral valve repair can also be safely performed in patients with mitral valve lesions as complications of acute and chronic endocarditis, in agreement with the findings of Muehrcke and colleagues[117,118] who advocated early intervention and repair in these cases to prevent leaflet destruction and vegetation embolization, and to preserve left ventricular function. In this small series, we have shown that mitral valve repair in children with infective endocarditis is safe, with no operative and long-term mortality. All patients had good functional status during long-term follow-up and not one had undergone mitral valve reoperation. The goals of surgery are to eradicate the focus of infection, to repair destroyed valve structures, to prevent the development of complications, and to prevent relapse of the infection. Mitral valve repair also has the advantage of preserving the subvalvular apparatus and ventricular function. As ventricular function is impaired at the time of operation, any preservation of function should achieve a reduction of operative mortality and improve postoperative remodeling and long-term survival.

Although infective endocarditis is an uncommon infection in the pediatric population, it is a serious problem once it occurs. Surgical treatment of infective endocarditis is necessary in patients who respond poorly to antibiotics, experience septic emboli, with annular abscess or hemodynamic instability and in those who are bacteriologically treated but in whom mitral insufficiency has developed as a result of valve destruction during infection.

Reconstruction of the mitral valve is a widely accepted surgical concept,[117-119] and has become a standard technique in patients with endocarditis only during recent years. Among the first to present data relating to successful mitral valve reconstruction in patients with acute endocarditis were Dreyfus and colleagues[118] who recommended early surgical intervention. In a study of 22 patients with active and healed endocarditis, Pagani and associates[119] found excellent early postoperative findings with respect to function and survival. Hendren et al. [120] reported that mitral valve reconstruction was an attractive alternative to replacement. Both Carpentier[34] and Dreyfuss[118] recommended aggressive

surgical intervention in infective valve disease consisting of debridement of the infected valve at an early stage while the infection is still limited to the heart valve tissue and before impairment of ventricular function occurs. They believed that the preservation of ventricular function is of primary importance and may be an adjunct in reducing operative mortality and morbidity by preserving functional status.

Annuloplasty is recommended when the mitral annulus is dilated to obtain a good surface of coaptation between the two leaflets. We refrain from using a prosthetic ring to support the annuloplasty as much as possible. Instead we used an autologous pericardial strip to reinforce the annulus or as pledgets for suture reinforcement, which had two distinct advantages – firstly that no prosthetic material was introduced into the infective operating field, and secondly that the need for anticoagulation was obviated. As there was no recurrence of endocarditis in our series, the advantage of using autologous pericardium is highly convincing. In complex mitral valve repair, its use also attenuates the mechanical stress on the suture lines. The most complex repairs were in 6 patients who underwent extensive resections of both leaflets with commissural reconstruction and papillary muscle division and reimplantation as well as chordal shortening. In most centers, these valves would have been replaced.

With absence of endocarditis recurrence and total freedom from any reoperation in our series, the advantage of using different valve reconstruction techniques (Kay Wooler and Paneth annuloplasty, modified Paneth-Hetzer posterior annuloplasty technique) as indicated, as well as careful reconstruction of the damaged leaflet and subvalvular apparatus, meticulous debridement and removal of vegetations, is evident. No complications comparable to those reported in other series [120-121] were observed. Since mitral valve repair preserves viable native tissues that are more resistant to infection than prosthetic material, this may explain our excellent postoperative outcome.

We recommend prompt surgical treatment once it is confirned that mitral valve is affected by infective endocarditis, to prevent further valvar damage, since native valve endocarditis begins on the native leaflet and remains there for some time before extending to the surrounding tissue.

Mitral valve repair in children with infective endocarditis can be performed without morbidity or mortality. This surgical therapy offers optimal ventricular remodeling due to preservation of the valvular and subvalvular apparatus, absence of reinfection, and in general, a lack of any anticoagulation therapy requirement. Long-term follow-up shows that functional improvement and follow-up echocardiography evaluating the degree of mitral valve regurgitation and stenosis are highly satisfactory. No patients were reoperated upon for the same mitral valve pathology or for any

other valve problems for that matter, indicating that the surgical techniques we employed were suitable to produce a satisfactory, long-lasting solution. We believe that our small collection of patients seen over 16 years in whom mitral valve repair was performed will be helpful in guiding clinical perspectives in children with infective endocarditis.

4.5 Surgical techniques and operative outcome of mitral valve repair in in lesion associated with a primary congenital heart defect (atrioventricular septal defect and a small left ventricle)

We have shown that small left ventricle in CAVSD may not necessarily be absolute and that the left ventricle has the potential to increase to a size that is sufficient to support the systemic circulation. In this regard, it was surmised that the volume-loaded right ventricle compresses the already small left ventricle, whereas after surgical correction of the CAVSD, primarily because of reversal of septal deviation, the left ventricle is allowed to attain its full status as a ventricle. Preoperative establishment of left ventricular size is definitely required to decide whether univentricular or biventricular repair is indicated. The key parameter, the long-axis ratio (LAR), is independent of septal deviation; hence, it is not only simple, but may also be far better than volumetry with its complex measurements.

Skillful surgical maneuver is a beneficial factor in the increase in LV size, as also reported by van Soon and colleagues.[122] The volume of the left ventricle was increased surgically by attachment of the ventricular septal patch a little more to the right of the ventricular crest than is usually done in the repair of CAVSD, thus contributing to the adequacy in LV size after operation. Precision must be exerted in the sizing of the patch, as oversizing results in patch redundancy with the potential for left AV valve regurgitation. It is our protocol to close the cleft whenever possible. The most controversial issue in the surgical treatment of CAVSD is the necessity of closing the mitral cleft.[66] Although several studies have reported that closure of the cleft can lead to stenosis of the left AV valve orifice, this has not been demonstrated in our series. This was probably the case because an age-related minimal normal valve diameter was used as a guide in all cases in cases during cleft closure to prevent valve stenosis.[66]. In fact, Alexi and colleagues[67] in our institution and others stated in their previous study that incidence of left-sided AV valve insufficiency seemed to be higher in patients with an initially unsutured cleft. Thus, we believe that the low incidence of late left-sided AV valve insufficiency maybe attributed to routinely suturing the cleft up to the minimal valve diameter and transecting the residual atrial septum.

Avoiding placement of sutures on the crura of the left-sided AV valves during closure of the ASD increases valve surface area and prevent leaflet puckering, thus improving leaflet coaptation. Fraisse and colleagues [65] have done an extensive study on the pathogenetic features and management of cleft

MV, and stated that cleft mitral valve may represent a "forme fruste" of an AV septal defect. They further stated that in patients with mitral regurgitation and AVSD, the smaller size of the mural leaflet along with two closely-spaced papillary muscles, may complicate repair and inaccurate cleft approximation may result postoperatively in significant mitral regurgitation or stenosis after insufficient or excessive closure.

The principal determinant of successful biventricular repair of CAVSD with small left ventricle appears to be related to the competence of the left AV valve and LV size. Indeed, at a median follow-up of 58.6 months, 13 patients had no left AV valve regurgitation, 3 had mild regurgitation; and no patient had any restriction of inflow or outflow. Significant left AV valve regurgitation is an indication for early reoperation. Remarkably, most often, the mitral valve can still be repaired. [67, 123] Many authors reported that management of left-sided AV valve in the repair of complete atrioventricular septal defects remains an area of controversy. Alexi and colleagues[67] reported that there are several factors contrbuting to left AV valve regurgitation besides a cleft that was left open during the operation or a dehiscence of cleft sutures, such as isolated dilatation of the valvar ring, severe valve deformity, dysplastic AV valve with marked loss of leaflet tissue and double orifice left AV valve. The incidence of early reoperation for regurgitant left AV (mitral) valve is 2-12%.[123] We confirm in our series that severe regurgitation of the left AV valve is the most important risk factor for reoperation.

4.7 Modified surgical techniques and long-term outcome of mitral valve repair

Because of a high prevalence of cardiac anomalies associated with mitral valve lesions in infants and children, whether the mitral valve should be repaired or replaced has been a matter of controversy. It is immensely formidable to reconstruct mitral valves in infants and children, not only because of their size, but primarily, because their mitral valves have immature and fragile leaflets, notwithstanding the associated congenital cardiac anomalies. Growth of these valves along with their somatic growth is another utmost concern. We encountered mitral valves significantly lacking in valve tissues, or severely dysplastic, with severe deformation of the subvalvular apparatus that rendered reconstruction impossible. Attempts to reconstruct and preserve these valves proved futile, since satisfactory functional results cannot be achieved; hence their valves were replaced during the same surgical procedure. Based on this experience, we learned that the main goal of surgical repair should be to achieve a satisfactory, if not ideal, MV function, rather than an ideal anatomical or morphological reconstruction.

We believe that the key to a satisfactory postoperative outcome is thorough preoperative evaluation and understanding of the valve abnormality with meticulous attention paid to the anatomical and

functional features of the MV apparatus and the precise mechanisms causing stenosis or insufficiency. Attempts to preserve the native mitral valve should always be encouraged, especially in infants and young children. Mitral valve repair offers the advantages of avoiding thromboembolism, preserving chordal and subvalvular apparatus function, and making reoperation unnecessary.

In this population, annular dilatation and prolapsed leaflet were frequently present. In all cases we tried to avoid placement of a rigid ring prosthesis for stabilization or to correct annulus abnormalities because of concerns about anticoagulation, subsequent somatic growth problems and the risk of a rigid prosthesis causing distortion of the heart cavities and/or contributing to left ventricular outflow tract obstruction.[124,125] Several studies consider ring annuloplasty for mitral valve incompetence obligatory in children over 2 years of age. [69,72,126,127] This concept is supported by their experience of a 25% incidence rate of significant residual mitral regurgitation after repair without ring insertion.[69] Other groups demonstrated that other types of annuloplasty techniques can be employed successfully in children and that prosthetic rings are not indispensable for achieving favorable results.[68,70] We do not use annuloplasty rings. In most of our patients, we employed the commissure plication annuloplasty technique to correct the annular dilatation, which in itself was reported to yield adequate long-term functional results.[128] In our institution, posterior annuloplasty using the Kay-Wooler technique, modified Paneth–Hetzer posterior annuloplasty and modified Gerbode-Hetzer plication plasty with reinforcement by an autologous pericardial strip proved to be highly satisfactory.

We found that the hammock mitral valve, defined as very dysplastic, with shortened chordae almost directly inserted in a muscular mass of the posterior wall of the left ventricle, thereby resulting in tethering of both leaflets, was one of the most challenging malformations to correct, as was also stated by other authors. [20,61,68,71,129,130] Hammock MV are stenotic; however, MI was also seen in most of our cases. The key to successful reconstruction was mobilization of these shortened chordae by splitting or incising them off the posterior muscular ventricular wall.

An adequate repair becomes a balance between residual stenosis and induced insufficiency, which is assessed intraoperatively with transesophageal echocardiography.

Few reports on MV repair in children have identified predictors for poor outcome in this group of patients, probably because of the small numbers of patients in each report. We have identified age $\leq$3 months, urgency of operation, concomitant procedures, operative times and valve status on discharge as risk factors for postoperative mortality and reoperation.

A major setback of MV reconstruction in children is durability of repair. Our actuarial survival and freedom from reoperation at 10 and 19 years are encouraging, especially in this population where 30% of patients were under 2 years of age. Its success is determined largely by intraoperative assessment of valve morphology and by the reconstruction techniques. We have continued to modify our surgical techniques to optimize our postoperative results.

4.8 Mitral valve growth after mitral valve repair

In infants and children, annuloplasty using untreated autologous pericardial strip to correct annular dilatation and to support any valve repair technique performed for mitral insufficiency has not only proven to stabilize the posterior annulus, but most of all allowed the growth of the mitral valve complex over time.[81] There is no exclusive report in the literature on posterior annulus stabilization with autologous pericardial strip. Autologous pericardium is not only costless and readily available with simple preparation in the operating theatre, but its flexibility maintains the physiologic movement of mitral annulus. It can be applied to the posterior annulus, the main target of mitral annuloplasty, to correct the annular dilatation, to increase the leaflet coaptation, to reinforce the annulus sutures[130,131] and to prevent future annular dilatation. Unlike the reported complication of other prosthetic annuloplasty,[1] like left ventricular outflow tract obstruction due to systolic anterior motion, no one in our series exhibited such complication. We believe that confining the annuloplasty to the posterior portion of the annulus is effective in avoiding such complication.

Pericardium has been attractive to the cardiac surgeon for a long time. Its ready availability, its ease of handling and its pliability make it an obvious choice when a defect must be eliminated or corrected. We do not treat the pericardial strip with glutaraldehyde, which was reported to enhance the tissue durability and prevent calcification. In our instititution, we are convinced that without treating autologous pericardium makes it more flexible to maintain the physiologic movement of the mitral annulus, construct a soft reinforcement which conforms to the natural tri-dimensional geometry of the mitral annulus, offers long term durability, and absence of thrombogenicity and calcification, as evidenced during reoperations. Stabilization of the posterior annulus with untreated autologous pericardium has positive influence on left ventricular function while preserving the contractile properties of the mitral annulus. These benefits, in addition to the easily accomplished surgical technique, the effective functioning of the remodelled valve, the preservation of the natural shape of the valve and allowing the valve to grow as the patient grows older, make this technique a useful surgical tool for mitral valve reconstructive surgeries.

6.0 Summary and Conclusions

These cumulative studies described our entire surgical experience with congenital and acquired mitral valve anomalies in children with biventricular hearts and concordant connections inclusive of atrioventricular septal defects. These confirmed the diversity and severity of congenital MV anomalies, which characteristically involved multiple components of the mitral valve complex.

In our institution, MV reconstruction is the surgical technique of choice for any kind of mitral disease in childhood. We firmly believe that repair allows continuous somatic and valve growth, and delays or eliminates the need for future valve replacement and lifelong anticoagulation, and obviating the known complications of valve replacement which frequently requires subsequent reoperation to implant a larger prosthesis. It must be assumed that the majority of, if not all, valves repaired during childhood, will eventually have to be replaced at some time in life. The concept of repair in childhood primarily aimed at growth of the patient to an age when, if necessary, an adult size prosthesis can be implanted. Strong predictors for poor overall survival and freedom from reoperation are age less than 3 months, urgency of surgery, concomitant procedures.

Surgical repair for MI from MV lesions with restricted leaflet motion in infants and children was performed with various suture-repair techniques and strategies, with excellent functional results and low reoperation rate at >20 years' follow-up. These techniques, which restored the mitral competence with effective functioning of the remodeled valve, the preservation of the natural shape of the valve and at the same time allows the valve to grow as the patient grows older, are useful surgical armamentarium in MV reconstructive surgeries in infants and children.

ALRP in combination with septal myectomy is an effective and safe treatment for children and adolescents with HOCM. Long-term follow-up shows sustained absence of systolic anterior motion, attenuation of mitral regurgitation, sustained improvement in functional status, and absence or reduction of outflow tract obstruction. Because the present technique offers a broadening of the surgical possibilities, we believe that ALRP could become the preferred choice in persistent LVOT obstruction and SAM.

We have demonstrated that in Shone's anomaly, an aggressive functional repair approach to the mitral valve and relief of the left ventricular outflow tract obstruction lead to long-term event-free survival in these children. Repair strategy has been challenging especially in hypoplastic valves in infants. Outcomes in this population are related to the degree to which mitral stenosis can be relieved.

Mitral valve repair for children with infective endocarditis can be performed without morbidity or mortality. This surgical therapy offers optimal ventricular remodeling due to preservation of the valvular and subvalvular apparatus, absence of reinfection, and in general, a lack of any anticoagulation therapy requirement. Long-term follow-up shows that functional improvement and follow-up echocardiography evaluating the degree of mitral valve regurgitation and stenosis are highly satisfactory.

In this small series of children with CAVSD with small left ventricle, biventricular repair was successfully accomplished. Our study in this small series of children with CAVSD with small left ventricle, MV repair offered the long-term survivors minimal or no regurgitation of either valve. We also believe that the low incidence of late left-sided AV (mitral) valve insufficiency maybe attributed to routinely suturing the cleft up to the minimal valve diameter and transecting the residual atrial septum. We confirm that right AV valve regurgitation is not a cause of reoperation after repair of CAVSD.

Stabilization of the posterior annulus with pericardial strip prevents further posterior annular dilation and allows anterior MV leaflet and its annulus to grow in relation to body size over time, as well as preserving flexible properties of the MV orifice.

6.0 Further studies

Repair strategies in floppy mitral valve in Marfan's syndrome, remodelling techniques in parachute and hammock valve and repair techniques in congenital mitral dysplasia are very interesting studies, which we have embarked upon. These studies will be presented in several future international meetings, and will be eventually published.

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8.0 Acknowledgment

My heartfelt and sincerest gratitude goes to **Prof. Dr.med Dr. h.c.mult. Roland Hetzer**, whose overwhelming and continued support through the years of my cardiothoracic surgical training and as a staff at Deutsches Herzzentrum Berlin, has given me a great motivation in pursuing advancement and excellence in my profession. Since the time I came to DHZB six years ago, he never fails to amaze me. I profited greatly from his surgical expertise and from his great knowledge in cardiac surgery. In so many ways, I profited a lot from his name alone. It is such an honor to have him as my surgical mentor. He is a living legend in the profession, and I feel very fortunate to have met him.

Dr. Norbert Franz has been always behind in most steps I made to advance my career.

I greatly appreciate the assistance of: **Ms. Christine Detschades, Ms. Daniela Moeske-Scholz** and **Ms. Heike Schultz** for collecting data and follow-up of patients; **Ms. Julia Stein** for being very helpful in data analysis; **Ms. Astrid Benhennour**, for providing me infinite literature; **Ms. Anne Gale** for her excellent editorial assistance; and **Mr. Helge Haselbach** and **Ms. Carla Weber**, for their graphic and design expertise.

I dedicate this dissertation to my son **Mariano**, whose birth has graced my training in general surgery and while growing up, trained himself to be a fine adolescent while I trained to become a cardiac surgeon. His dreams of eventually going to this specialty has not been hampered by my frequent absence and the daily stresses and great demands of this profession, which he absorbed gracefully without complaints, having regarded it as a normal way of living.

9.0 Statutory Declaration

ERKLÄRUNG

§ $ Abs:3 (k) der HabOMed der Charite

Hiermit erkläre ich, dass

- weder früher noch gleichseitig ein Habilitationsverfahren durchgeführt oder angemeldet wurde,
- die vorgelegte Habilitationsschrift ohne fremde Hilfe verfasst, die beschriebenen Ergebnisse selbst gewonnen sowie die verwendeten Hilfsmittel, die Zusammenarbeit mit anderen Wissenschaftlern/Wissenschaftslerinnen und mit technischen Hilfskräften sowie die verwendete Literatur vollstaändig in der Habilitationsschirft angegeben wurden.
- mir die geltende Habilitationsordnung bekannt ist.

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