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Assessment:

When on my first mentorship visit with Dr. Timothy Pirolli we discussed my involvement in a research on mitral valvuloplasty and mitral valve replacement in infants less than one year old, and in doing so I was given a series of articles to annotate and research. Knowing that most of this information required background knowledge I have yet to attain Dr. Pirolli gave me an article that covers congenital malformations of the mitral valve. By annotating this article and creating this research assessment I will gain the background knowledge to better understand the articles I will be using during our research.

The article begins with the normal anatomy of the mitral valve and understanding the normal anatomy of the mitral valve will lead to a deeper understanding of its malformations. My prior knowledge of the anatomy mitral valve was limited to none since the anatomy of the heart was never deeply taught upon in my high school level science classes. I also have never learned about valve specific anatomy and I now realize that my knowledge of the anatomy of the heart is

something worth researching upon to better understand the congenital heart defects dealt with in pediatric cardiothoracic surgery. The mitral valve consists of the annulus which C shaped like a saddle. The mitral valve consists of two leaflets the anterior (A1, A2, and A3) and the posterior leaflet (P1, P2, and P3) and between the two leaflets are the lines of commissure, anterolateral and posteromedial. The mitral valve also consists of the chordae tendinae and papillary muscles. When all of this anatomy is working properly the mitral valve allows blood flow from the left atrium into the left ventricle, but not the other way. Now with the basic understanding of the correct anatomy of the mitral valve I can go onto understanding the congenital malformations.

The first discussed type of mitral valve malformation is mitral valve prolapse which occurs when the leaflets extend above the plane of the mitral annulus during ventricular systole. This is the most common cardiac vascular anomaly; however, its uncommon before adolescence. Treatment usually includes mitral valve repair/annuloplasty for severe mitral regurgitation or ventricular enlargement. Although a common malformation this malformation will probably not relate to our research since its uncommonly seen in infants or even children. The next discussed was the isolated cleft, a rare malformation, which is division of one of the leaflets and is divided within two groups with normal great arteries and with abnormal conus. When suffering from severe regurgitation mitral valve replacement patients need to be operated early in life, which means this malformation will possibly be related with our research. Next is a double orifice mitral valve which is extremely rare and seen only in 1% of autopsies of congenital heart defects and is rarely isolated and found within more complex cardiac malformations. Double orifice mitral valves contain a single fibrous annulus with two orifices opening into the left ventricle. It is divided by three types the incomplete bridge, the complete bridge, and hole type.

Isolated double orifice malformations require no repair, but when combined with other malformations surgery is often performed. This malformation is probably not related to our research since this defect is rarely found isolated and does not require surgery. A mitral ring was also briefly talked about, this an extremely rare defect divided into two parts the supramitral ring which is a fibrous membrane originating above the mitral annulus and the intramitral ring with is located beneath the orifice of the left atrial appendage. This defect is exceptionally isolated and is often combined with various other defects like the double orifice mitral valve. Being non isolated this malformation will probably be unrelated to our research. The next types of malformations relate to the tensor apparatus starting with an arcade/hammock valve. In this malformation the valve mimics a hammock when observed from the surgeon's perspective and happens when the tendinous cords are thickened and short, which leads to abnormal leaflet excursion which may cause stenosis or insufficiency. When not severe this malformation may have a late discovery and a surgical repair will happen later in life. I believe this malformation will be unrelated to my research.

The following part of the article talks of the management of these malformations, which is specifically related to our research. Often a surgery or intervention is rarely needed before the first year of life regarding mitral regurgitation; however, regarding mitral stenosis may require early surgery which is important to note when leading into more research for my publication. In age less than one often hammock mitral valve and associated cardiac anomalies are seen. Regarding malformations of the leaflets, chordae tendinae, and papillary muscles make early surgery extremely important which highlights the importance of our research. This article also highlights the importance of performing mitral valve repairs rather than replacements since

mechanical valves often lead to treatments and procedures to ensure the valve is working correctly creating a lifetime of valve control.

Overall, this article highlighted the debated results of intervention of the mitral valve and further encouraged me to follow through with this publication and research and gave me the background knowledge needed to continue to learn about mitral valvuloplasty and mitral valve repair. I now see the impact we could make on the field of pediatric cardiothoracic surgery by allowing the growth of information within intervention.



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REVIEW

Congenital malformations of the mitral valve

Malformations congénitales de la valve mitrale

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MOTS CLÉS

Cardiopathie congénitale ;
 Valve mitrale ;
 Prolapsus mitral ;
 Valve mitral en parachute ;
 Fente mitrale ;
 Échocardiographie

Summary Congenital malformations of the mitral valve may be encountered in isolation or in association with other congenital heart defects. Each level of the mitral valve complex may be affected, according to the embryological development, explaining the fact that these lesions are sometimes associated with each other. As a perfect preoperative assessment is of importance, good knowledge of both normal and abnormal anatomy is required in order to guide the surgeon accurately. This review presents the different embryological, anatomical and echocardiographic aspects of the congenital mitral anomalies.

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Résumé Les malformations congénitales de la valve mitrale peuvent être rencontrées isolément ou en association avec d'autres cardiopathies congénitales. Ainsi que le montre l'embryologie, chaque étage du complexe valvulaire mitral peut être atteint. Cela explique le fait que ces lésions peuvent parfois s'associer entre elles. Une parfaite évaluation préopératoire étant cruciale, une bonne connaissance de l'anatomie normale et des malformations est requise afin de pouvoir guider précisément le chirurgien dans son geste. Cette revue présente les différents aspects embryologiques, anatomiques et échocardiographiques des anomalies congénitales de la valve mitrale.

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Abbreviations: AVSD, atrioventricular septal defect; DOMV, double orifice mitral valve; MVP, mitral valve prolapse; PLAMV, parachute-like asymmetric mitral valve; PMV, parachute mitral valve; SMV, straddling mitral valve; TGF- β , transforming growth factor beta.

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research focus
 important to understand why bla. research

Why there is little on isolated

Background

Congenital anomalies of the mitral valve represent a wide spectrum of lesions that are often associated with other congenital heart anomalies. In an echocardiographic study, congenital malformations of the mitral valve were detected in almost 0.5% of the 13,400 subjects [1]. These lesions can have a variable impact on valve function. When necessary, surgical repair provides good long-term results [2-4]. Although mitral valve replacement appears to provide acceptable mid- and long-term results [5,6], mitral valve repair is always preferable when possible. Because suboptimal primary repair is a significant predictor for reoperation, the successful management of congenital mitral valve disease is closely dependent on the preoperative assessment of the anatomical substrate [7]. An accurate description of the malformations can be achieved through echocardiography but requires prior knowledge of these lesions. Thus, the mitral valve should be analysed as an entire complex, including the valvar leaflets, tensor apparatus and papillary muscles. This review will discuss the different congenital malformations that can affect the mitral valve.

mitral valve repair

The subvalvular apparatus is composed of chordae tendinae and papillary muscles (Fig. 1). Chordae tendinae connect all parts of the leaflets to two ventricular papillary muscles. Leaflet cords have several shapes and are attached to the leaflets at various sites. Thus, marginal cords (attached to the free edge), rough zone cords (attached to the rough zone) and strut cords (attached to the basal portion of the posterior leaflet) have been identified [10]. In most cases, papillary muscles are organized as two groups of closed papillary muscles as opposed to two distinct muscles, which arise from the apical and two thirds of the left ventricular wall. The tendinous cords extend from their tips. Papillary muscles are in anterolateral and posteromedial positions. All these structures can be analysed accurately by transthoracic echocardiography (Fig. 2).

Embryology

Mitral valve formation begins during the fourth week of gestation. Knowledge of its embryology is very useful for understanding the various anomalies that can affect it. During the sixth week, fusion of the endocardial cushions divides the atrioventricular canal into right and left atrioventricular junctions (Fig. 3) [9]. Failure of fusion of the superior and inferior cushions, presumably secondary to a deficiency of the vestibular spine, is responsible for producing AVSD. Normally, the lateral cushion forms the posterior mitral leaflet while the anterior leaflet derives from the apposition of the left part of the superior and inferior cushions. At the eighth week, the shape of the orifice looks like a crescent, the two ends of which are connected to compacting columns in the trabecular muscle of the left ventricle. These columns form a muscular ridge, the anterior and posterior parts of which become the papillary muscles [13]. The transformation of the ridge into papillary muscles implies a gradual loosening of muscle, which is called delamination (Fig. 3). The abnormal compaction of the ventricular trabecular myocardium is responsible for producing the PMV. Simultaneously, as for the tricuspid valve, the cushion tissue loses contact with the myocardium of the ridge, except at the insertion of the future tendinous cords. The very rare Ebstein's malformation of the mitral valve results from a failure of excavation of the posterior leaflet from the parietal ventricular wall. The chordae can be individualized between the eleventh and thirteenth week of development by the appearance of defects in the cushion tissue at the place where the tips of the papillary muscles are attached to the leaflets. As proved by their having the same immunohistochemical characteristics, both leaflets and chordae originate from the cushion tissue [13], whereas papillary muscles are derived from the ventricular myocardium. A lack of development of the tendinous cords results in hammock or arcade mitral valve. The more severe anomaly of the leaflet is represented by the imperforate mitral valve. Finally, as each stage of this embryological development may be abnormal, the different malformations of the mitral valve can be either isolated or associated.

subvalvular apparatus chordae tendinae papillary muscles

Normal anatomy

The mitral valve, so named because of its resemblance to the episcopal mitre, is bicuspid and marks the left atrioventricular junction. The mitral valve is better understood as a complex that comprises the annulus, the anterior and posterior leaflets, the chordae tendinae and the papillary muscles. The annulus is surgically defined as the level of visible transition between the left atrial myocardium and the whitish leaflet. According to its anatomical definition, it is considered as the fibrous hingeline of the valvar leaflets [8]. Due to the fibrous continuity between the aortic valve and the anterior (or aortic) mitral leaflet, defining its exact limits is extremely difficult [9]. The annulus is saddle-shaped (or D-shaped) (Fig. 1) [10]; it is a dynamic structure that contracts and reduces its size during systole [8].

ref. I not is knowledge nearby anatomy

mitral valve = valvar leaflets tensor apparatus papillary muscles

The mitral leaflets are uninterrupted structures, which vary in shape and in circumferential length. They are usually divided into anterior and posterior segments (Fig. 1). At present, many authors have separated them into aortic (anterior) and mural (posterior) leaflets because of their connection with the aortic valve and the posterior wall of the left ventricle. Unlike the tricuspid valve, the mitral valve leaflets have no attachments to the septum. During systole, when the leaflets meet to close the ventricle, the line of coaptation, also called the commissure, looks like a smile. The terms anterolateral and posteromedial commissures, sometimes used to designate each end of the closure line, are unsuitable because a bifoliate valve can have only one zone of apposition between the two leaflets [11]. Coaptation occurs along the leaflet edge in the rough zone. According to the classification by Carpentier et al. [12], the free edge of the posterior leaflet is divided into three scallops: P1 (lateral); P2 (middle); and P3 (medial). The anterior leaflet is subdivided into A1, A2 and A3 regions that are opposite the scallops of the posterior leaflet.

posterior leaflet (P1, P2, P3) anterior leaflet (A1, A2, A3)

subvalvular apparatus chordae tendinae papillary muscles

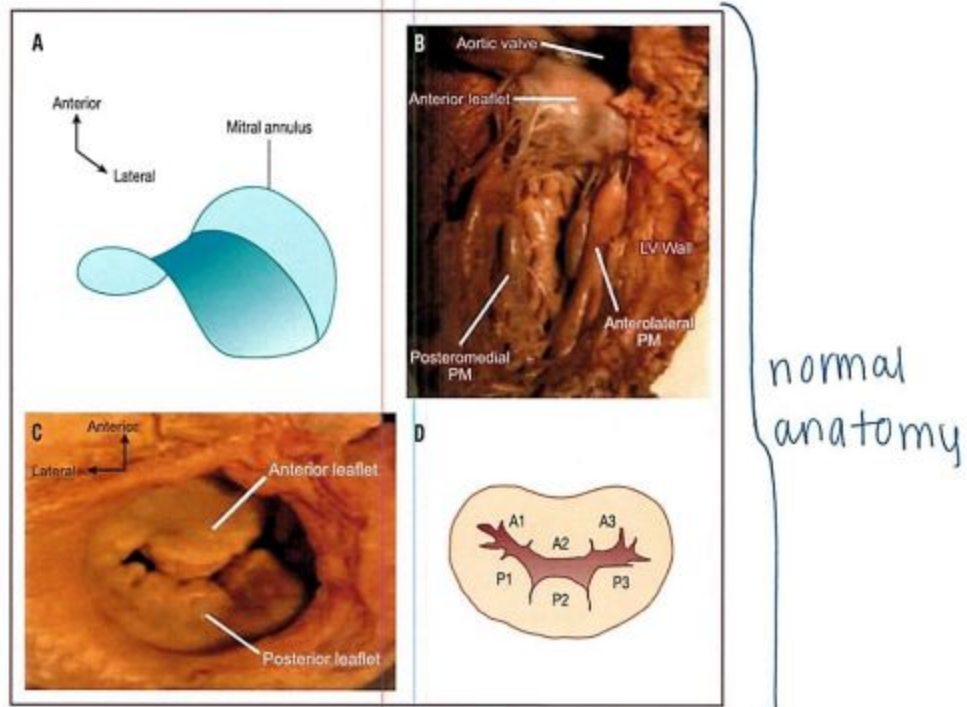


Figure 1. Normal mitral anatomy. (A) Schematic representation of the saddle-shaped mitral annulus. (B) Anatomical photograph of a normal mitral complex with its two papillary muscles connected to the leaflets by chordae tendinae. The aortic valve is in direct continuity with the anterior leaflet of the mitral valve. (C) Photograph of a normal mitral valve seen from the left atrium (as seen by a surgeon). (D) Both leaflets are divided into three scallops according to the classification by Carpentier et al. [12]. LV: left ventricle; PM: papillary muscle. Adapted from [12].

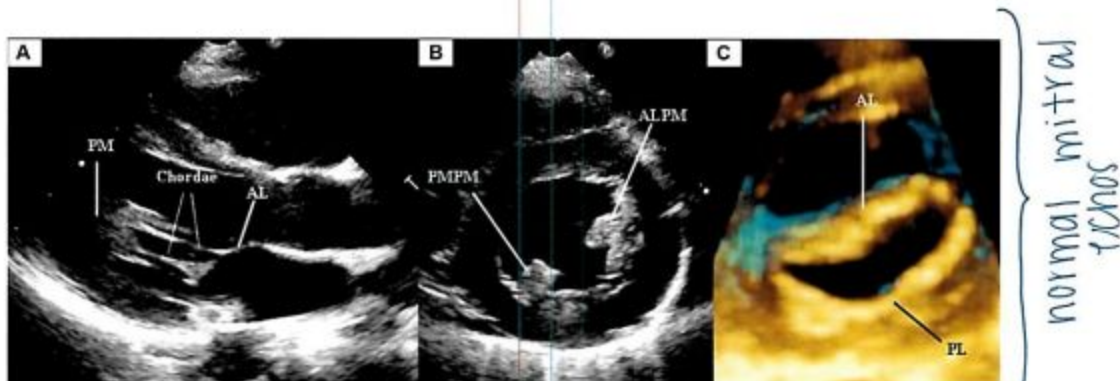


Figure 2. Normal mitral echocardiography. (A) Echocardiographic parasternal long-axis view showing a normal mitral complex. (B) Echocardiographic parasternal short-axis view showing the normal position of the papillary muscles. (C) Three-dimensional echocardiography of a normal mitral valve. AL: anterior leaflet; ALPM: anterolateral papillary muscle; PL: posterior leaflet; PM: papillary muscle; PMPM: posteromedial papillary muscle.

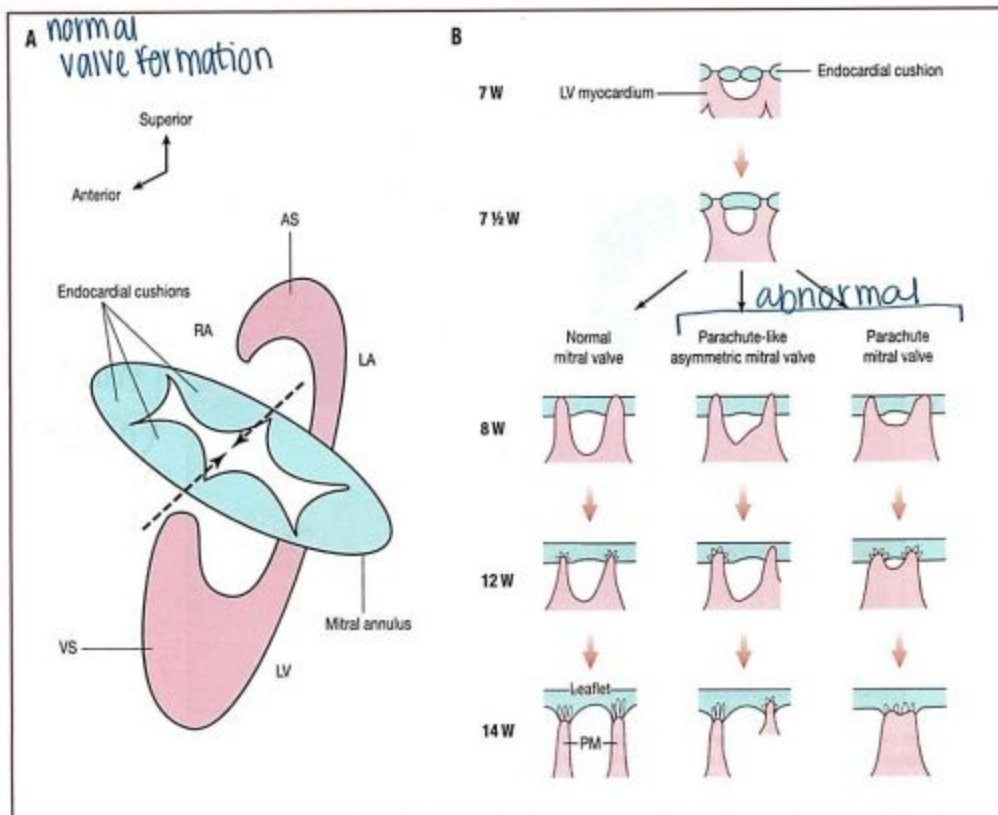


Figure 3. Mitral embryology. (A) Schematic representation of normal atrioventricular valve formation. The fusion of the superior and inferior endocardial cushions (arrows) will divide the atrioventricular canal into right and left atrioventricular junctions. (B) Schematic representation of normal and abnormal development of mitral papillary muscles. Normally, the progressive loosening of left ventricular muscle (myocardial delamination) results in the formation of two separate equal-sized papillary muscles. Both leaflets and chordae tendinae are derived from the endocardial cushions. Asymmetric papillary muscles develop when one of the two papillary muscles does not correctly delaminate from the left ventricular wall, with its tip remaining attached to the cushions. Abnormal compaction of the left ventricular myocardium is responsible for producing a true parachute mitral valve. AS: atrial septum; LA: left atrium; LV: left ventricle; PM: papillary muscles; RA: right atrium; VS: ventricular septum; W: week of gestation. Adapted from [13].

prob. won't relate to study
most common

Anomalies of the leaflets
Mitral valve prolapse

above plane

MVP occurs when the leaflets extend above the plane of the mitral annulus during ventricular systole. It is the most common cardiac valvular anomaly in developed countries. Myxomatous degeneration is the main aetiology of prolapsing valvar leaflets, explaining the fact that MVP is uncommon before adolescence. Indeed, the prevalence of MVP was 0.7% in a population of healthy teenagers [14]. In comparison, the Framingham study revealed that 2.4% of adult subjects had an MVP [15]. When MVP occurs during childhood, it generally integrates into a congenital disorder affecting the

connective tissue, such as Marfan syndrome, Ehler-Danlos syndrome, osteogenesis imperfecta, dominant cutis laxa or pseudoxanthoma elasticum.

As previously pointed out, the mitral valvar annulus is not perfectly circular but appears more like a saddle that has high and low points. The high points are represented by the anterior and posterior parts of the annulus, while the medial and lateral parts correspond to the low points. This particular morphology explains the fact that, in the past, MVP was broadly overestimated. Indeed, the normal leaflets can falsely appear to prolapse in certain echocardiographic views, especially in the apical two- and four-chamber views. New echocardiographic criteria have consequently been established based on the understanding



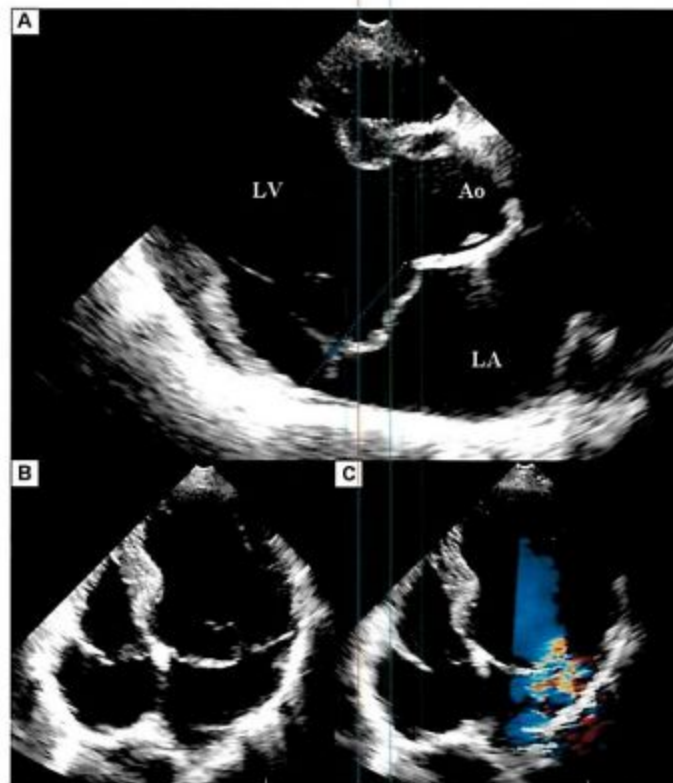


Figure 4. Mitral valve prolapse. (A) Echocardiographic parasternal long-axis view showing the mitral leaflets prolapsing more than 2 mm above the plane of the mitral annulus (dotted line) during systole in a child with Marfan syndrome. (B) Echocardiographic apical four-chamber view showing bileaflet prolapse in the same patient. (C) Colour Doppler view showing moderate mitral regurgitation. Ao: aorta; LA: left atrium; LV: left ventricle.

of the three-dimensional non-planar shape of the mitral annulus. Since then, echographical MVP has been defined as a single or bileaflet prolapse located at least 2 mm beyond the long-axis annular plane, with or without a thickening of leaflets (Fig. 4) [16]. It has been clearly proven that only prolapses shown in the parasternal long-axis view do not satisfy the diagnosis [17]. A classic prolapse is defined as a leaflet thickening exceeding 5 mm, whereas a prolapse with a lesser degree of leaflet thickening is referred to as non-classic.

In children, MVP may be secondary to a distortion of the left ventricular geometry, as seen in unrepaired atrial septal defects (right ventricular volume overloading and left ventricular size reduction). In this case, the mitral valve is histologically normal and the prolapse usually resolves postoperatively. MVP is also observed in cases of connective tissue disorders [16]. The percentage of MVPs associated with Marfan syndrome ranges from 40 to 91% [16,18]. Marfan

syndrome is associated with mutations in fibrillin-1 on chromosome 15q21.1 and with mutations in TGF- β receptor 2 on chromosome 3p24.2-p25 [19]. Fibrillin-1 is involved in the activation of TGF- β . Several studies have suggested that abnormalities in the TGF- β signalling pathway represent a common pathway for the development of the Marfan phenotype. It is a diffuse disease process, probably due to structural protein defects in cardiac tissues (fibrillin 1), which explains the concomitant illness of the aortic root and mitral valve. MVP most commonly involves both leaflets and is symmetrical in Marfan syndrome, whereas it more frequently affects one leaflet (posterior) in myxomatous degeneration [18].

The most serious complication is severe mitral valve regurgitation, although it is uncommon [20]. Vasodilator therapy is not recommended for the treatment of asymptomatic patients with severe mitral regurgitation and normal left ventricular function, as this may increase the risk of paradoxical worsening in mitral regurgitation [16].

Mitral valve repair is recommended in patients with symptomatic severe mitral regurgitation or in asymptomatic patients with ventricular enlargement or dysfunction. Surgical technique consists of resection of the prolapsed part of the leaflet, with or without an annuloplasty. The risk of endocarditis is higher for patients with MVP than for the general population, especially if the valve has thickened leaflets [16], but antibiotic prophylaxis is not strictly recommended according to the current American College of Cardiology/American Heart Association guidelines [21].

Isolated cleft

Isolated cleft of the anterior mitral valve leaflet is a rare but well-known finding, the origin of which is under debate. Indeed, some authors have considered isolated cleft to be a 'forme fruste' of AVSD whereas others have supposed it to be a distinct morphological entity. The definition of a mitral cleft is a division of one of the leaflets (usually the anterior leaflet) of the mitral valve. This must not be mistaken with the so-called 'cleft' in AVSD [22]. AVSD is characterized by a five-leaflet valve guarding a common atrioventricular junction: superior bridging leaflet; inferior bridging leaflet; left mural leaflet; right inferior leaflet; and right anterosuperior leaflet [22]. AVSD can be separated into complete and partial forms, depending on the degree of attachment of the superior and inferior bridging leaflets to the crest of the ventricular septum and to the inferior rim of the atrial septum. In complete AVSD, there is a single common orifice. The partial form is also defined by a common valve annulus but with the existence of two separate orifices due to a tongue of tissue joining the free margins of the superior and inferior bridging leaflets [23]. A characteristic finding of AVSD is the shorter inlet dimension of the left ventricular septal surface compared with its outlet dimension, whereas in a normal heart, inlet and outlet lengths are nearly equal. AVSD is believed to be the consequence of a deficiency in the development of the vestibular spine. In their large autopsic series, Van Praagh et al. stated that isolated cleft may be classified into two distinct groups: cleft with normally related great arteries, which would be a milder variation of the abnormal development of the atrioventricular canal; and cleft with abnormal conus associated with transposition of the great arteries or double outlet right ventricle [24]. Supporting the hypothesis of a common origin with AVSD, another series reported cases of isolated clefts with intact septal structures but with characteristics of AVSD [23]. Opposing this theory, some surgical studies did not find any feature of AVSD, such as the position of the papillary muscle, in all cases of isolated mitral cleft [25,26]. Kohl et al. clearly demonstrated that in AVSD, the positions of both papillary muscles were rotated counterclockwise (Fig. 5), whereas in isolated cleft, the position of the papillary muscles was similar to that in normal children [27]. Indeed, in AVSD, the posteromedial papillary muscle is more rotated than the anterolateral one, making it a good marker of this lesion. Moreover, in AVSD, the cleft points towards the ventricular inlet septum, whereas in isolated cleft, it is usually more directed towards the aortic root (Fig. 5). On transthoracic echocardiography, it looks like a slit-like hole in the anterior

mitral leaflet (Fig. 6). Chordal attachments may connect the edges of the cleft to the ventricular septum and subsequently create a subaortic obstruction [23]. More rarely, isolated cleft may be seen in the posterior leaflet of the mitral valve (Fig. 6) [28]. Although it may occur at any segment of the posterior leaflet, the predominant localization of the cleft is within scallop P2 [29]. Cleft of the posterior mitral leaflet has been reported in association with counterclockwise malrotation of the papillary muscles that may, again, lead one to suspect a common embryological origin with AVSD [30]. Mitral regurgitation, which is severe in 50% of cases, seems to be well analysed using three-dimensional echocardiography [31].

Mitral valve repair of isolated cleft associated with mitral regurgitation is preferred to mitral valve replacement and usually consists of a direct suture of the cleft [25,32]. Because of progression of the mitral regurgitation, patients may be operated on early in life [32]. When surgical treatment is performed in adults, the edges of the leaflets tend to be thicker and more retracted [33], which makes the repair more complicated, requiring interposition of patches on the mitral valve [25].

Double orifice mitral valve

DOMV is a rare condition occurring in 1% of autopsied cases of congenital heart disease [34]. DOMV is rarely isolated but usually an ancillary finding in the setting of a more complex congenital cardiac anomaly [35]. This lesion is usually found in association with AVSD (52%), obstructive left-sided lesions (41%) and cyanotic heart disease. Several cases of DOMV were also reported in association with non-compaction of the left ventricle [36–38]. DOMV is defined as a single fibrous annulus with two orifices opening into the left ventricle (Fig. 7). It differs from duplicate mitral valve, which is defined as two mitral valve annuli and valves, each with its own set of leaflets, commissures, chordae and papillary muscles. DOMV must also be distinguished from an acquired defect after mitral surgery. According to Trowitzsch et al. [39], DOMV is usually classified into three types: the 'incomplete bridge type' is characterized by a small strand of tissue connecting the anterior and posterior leaflets at the leaflet edge level; in the 'complete bridge type', a fibrous bridge divides the atrioventricular orifice completely from the leaflet edge all the way through the valve annulus; finally, in the 'hole type' (eccentric), a secondary orifice with subvalvular apparatus occurs in the lateral commissure of the mitral valve. In their autopsic series, Baño-Rodrigo et al. found consistently an anomaly of the tensor apparatus [34]. The two orifices are of equal size in 15% of cases, while a smaller (accessory) posteromedial orifice is encountered in 44% of cases.

Because there are no unusual signs to suggest DOMV, the clinical presentation is variable, mainly depending on the associated cardiac lesion. Symptoms are related to the degree of mitral insufficiency and/or stenosis. Mitral insufficiency occurred in 43% of cases, mitral stenosis in 13% and both stenosis and insufficiency in 6.5%. There is no functional consequence of DOMV in 37% of cases [35]. Transthoracic echocardiography is efficient for diagnosing and evaluating DOMV. The two distinct orifices are clearly recognized in parasternal short-axis views (Fig. 7). Rather than the

- Two groups
1. w/ normal great arteries
 2. w/ abnormal conus

- Three types
1. the incomplete bridge
 2. the complete bridge
 3. hole type

severe

may relate to study

may not relate

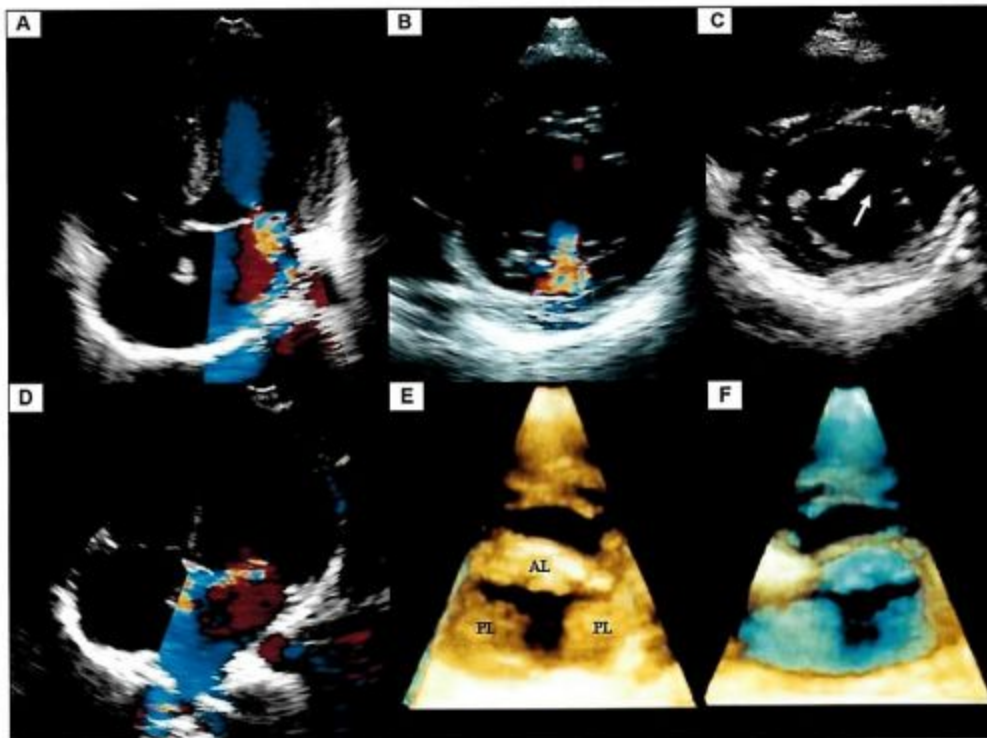


Figure 6. Echocardiographic comparison of isolated anterior mitral cleft and isolated posterior mitral cleft. (A) Two-dimensional echocardiographic apical four-chamber view showing the eccentric mitral regurgitation of an isolated anterior mitral cleft. The regurgitation jet is passing along the lateral wall of the left atrium. Parasternal short-axis view showing (B) mitral regurgitation in colour Doppler mode and (C) the cleft, which looks like a slit-like hole, pointing toward the aortic root (white arrow). (D) Two-dimensional echocardiographic apical four-chamber view showing the eccentric mitral regurgitation of an isolated posterior mitral cleft. The regurgitation jet is passing along the atrial septum. (E and F) Three-dimensional echocardiographic views of the posterior mitral cleft separating the posterior leaflet into two equal parts. AL: anterior leaflet; PL: posterior leaflet.

must be distinguished from cor triatriatum sinister, which is a fibromuscular membrane, clearly separated from the mitral valve (proximal to the left atrial appendage) that divides the left atrium into two parts. Cor triatriatum sinister is believed to be the consequence of a failure in the embryological development of the common pulmonary vein, while the two types of mitral ring might still have different embryological origins. Indeed, the intramitral ring seems to be a part of an intrinsic mitral disease, whereas the supramitral type is more like an obstruction of the left atrial outlet. Nevertheless, the supramitral ring may be described as a valvar lesion rather than supralvalvar because the annulus is an integral part of the mitral valve [44]. The ring can be either complete, circumferential or partial. It creates a stenosis that is usually progressive with a median age at diagnosis of 36 months in the largest published series [45]. Patients usually present with clinical features of congestive heart failure. Transthoracic echocardiography accurately detects

the mitral ring in up to 70% of cases [43]. Postoperative outcome is better for supramitral ring, with no need for reoperation after the ring excision, compared with frequent recurrence (50%) in case of intramitral ring [45]. In such cases, concomitant surgery of the tensor apparatus must often be performed to obtain sufficient haemodynamics.

Ebstein's malformation of the mitral valve

Ebstein's malformation of the left-sided atrioventricular valve has been reported a few times in cases of corrected transposition of the great arteries [46], but, in this situation, the involved valve was obviously of tricuspid morphology. The first case of Ebstein's malformation of a morphological mitral valve was described in 1976 by Ruschhaupt et al. [47]. The malformation exclusively affects the posterior valve leaflet, which is plastered

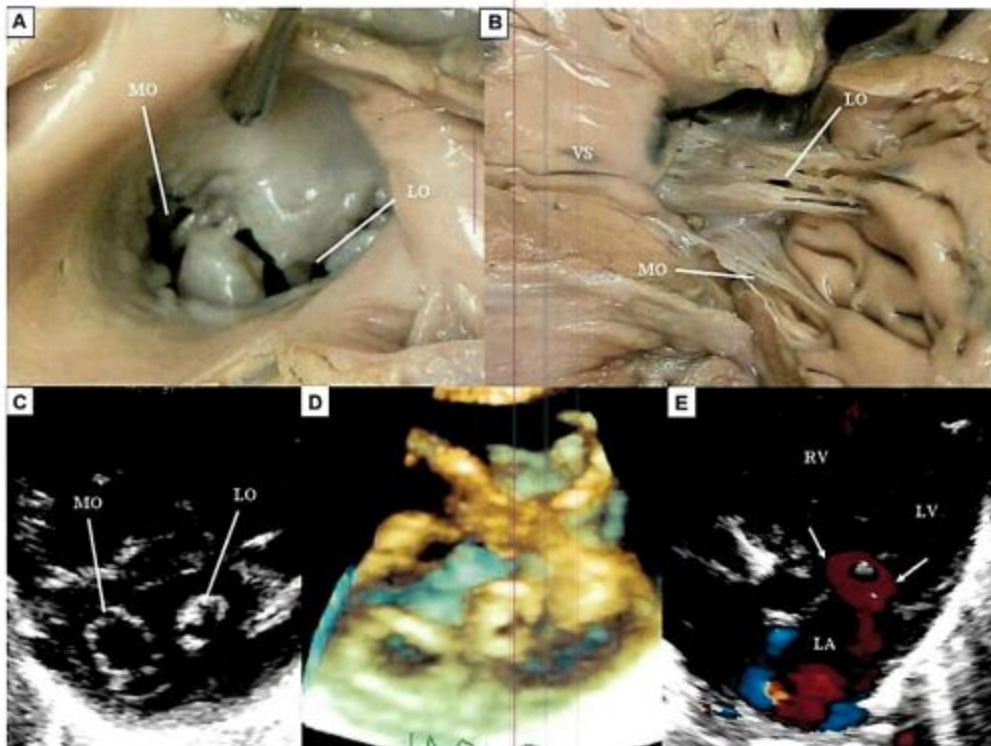


Figure 7. Double orifice mitral valve. (A) Photograph of a double orifice mitral valve seen by the left atrium (as seen by a surgeon), with a single fibrous orifice and (B) a double orifice mitral valve associated with partial atrioventricular septal defect seen by the left ventricle. (C, D) Two-dimensional and three-dimensional echocardiographic parasternal short-axis views showing the two distinct orifices. (E) Apical four-chamber Doppler colour view showing two typical anterograde flows (arrows) through the mitral valve. LA: left atrium; LO: lateral orifice; LV: left ventricle; MO: medial orifice; RV: right ventricle; VS: ventricular septum.

into the left ventricle wall, thus displacing the mitral valve orifice downward into the left ventricle. Unlike Ebstein's malformation of the tricuspid valve, the atrialized inlet portion is usually not thinned [48]. This exceedingly rare anatomical condition causes mitral insufficiency.

prob. will not be included

Anomalies of the tensor apparatus

Arcade or hammock valve

Anomalous mitral arcade was first described as a direct connection of the papillary muscles to the mitral leaflets, either directly or through the interposition of unusually short chordae [49]. This congenital malformation of the tensor apparatus is sometimes called hammock valve because it mimics a hammock when the valve is observed from an atrial aspect (as seen by a surgeon) (Fig. 9). The tendinous cords are thickened and extremely short, thus reducing the intercordal spaces and leading to an abnormal excursion

of the leaflets that may cause both stenosis and insufficiency. When the space between the abnormal chordae is completely obliterated, a fibrous (muscular) bridge (band) joins the two papillary muscles (Fig. 9). In the most severe form, with no chordae tendinae at all, the papillary muscles are directly fused with the free edge of the leaflet. Although mitral arcade is not an anomaly of the papillary muscles, it may be seen in association with PMV. This malformation is believed to be the result of an arrest in the developmental stage of the mitral valve before attenuation and lengthening of the collagenized chordae tendinae [49]. Echocardiographical appearance shows the short chordae and restricted motion of the leaflets with limited coaptation but also, in Doppler colour mode, multiple jets through the reduced intercordal spaces (Fig. 9) [11]. Mitral regurgitation progressively gets worse, with or without concomitant stenosis. However, the valve may function relatively normally for many years, as shown by late discoveries [50]. When necessary, conservative surgery will create two separated papillary muscles by resection of the muscular band [51].

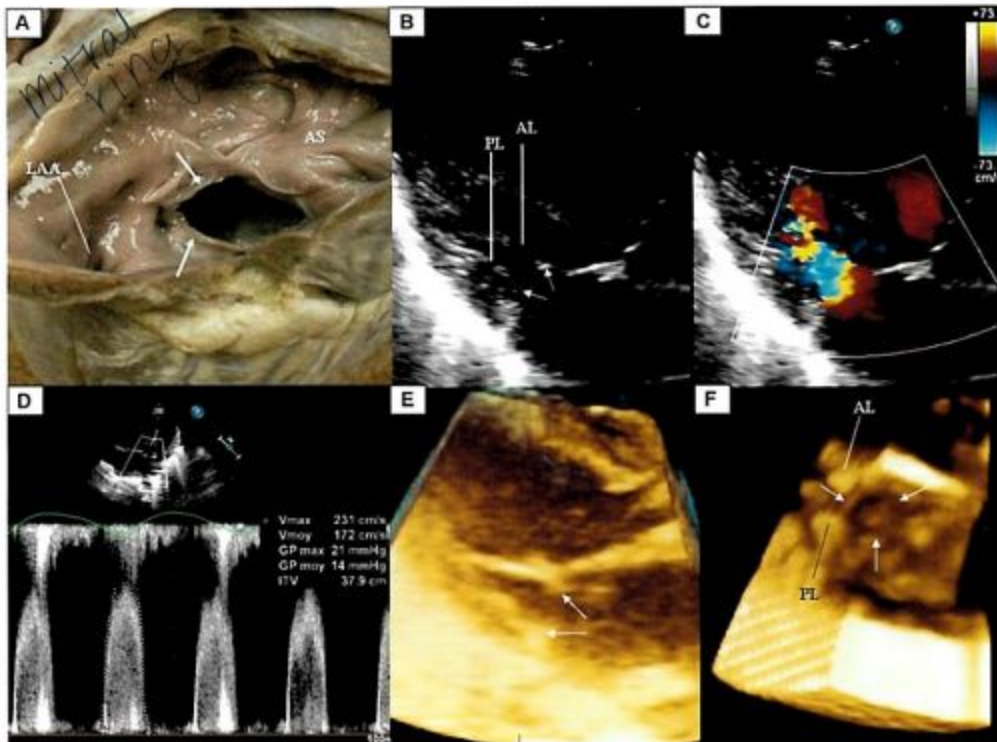


Figure 8. Mitral ring. (A) Photograph showing a supramitral ring (arrows) seen from the left atrium. The membrane is originating just above the mitral annulus, beneath the orifice of the left atrial appendage. (B) Two-dimensional echocardiographic parasternal long-axis view showing an intramitral ring (arrows) located within the funnel created by the mitral leaflets and (C) Doppler colour mode showing blood flow acceleration that begins at the insertion of the membrane. (D) Transmittal pulsed Doppler acquisition showing mitral stenosis. (E) Three-dimensional echocardiographic parasternal long-axis view showing the same intramitral ring (arrows). (F) Three-dimensional view from the left atrium. AL: anterior leaflet; AS: atrial septum; LAA: left atrial appendage; PL: posterior leaflet.

Straddling mitral valve

SMV is defined by an abnormal attachment of the mitral chordae to both ventricles [52]. SMV is consequently always associated with a ventricular septal defect. According to this definition, an AVSD nearly always straddles, but the term 'straddling' can only be applied to true mitral or tricuspid valves. The mitral valve always straddles through a conoventricular (misalignment) type of ventricular septal defect. SMV is almost always associated with conotruncal anomalies, such as double outlet right ventricle (Fig. 10) or transposition of the great arteries [53]. SMV must be distinguished from the overriding of the mitral valve, which qualifies a mitral annulus committed to the two ventricular chambers. In that case, the mitral valve is shared between the ventricles [52]. A mitral valve can straddle and/or override [54]. Surgical management of SMV is closely dependent on the more complex associated cardiac anomaly.

Anomalies of the papillary muscles

Parachute mitral valve *Common*

Among the causes of congenital mitral stenosis, PMV is frequently encountered, as shown by the incidence of 0.17% reported in a community echocardiographic study [1]. True PMV is characterized by unifocal attachment of the mitral valve chordae to a single (or fused) papillary muscle. This single papillary muscle is usually centrally placed and receives all chordae from both mitral valve leaflets (Fig. 11). In PLAMV, chordae are distributed unequally between two identifiable papillary muscles, with most or all of the chordae converging on a dominant papillary muscle [13]. The dominant papillary muscle, classically posteromedial [55], is of normal size, whereas the other is elongated and displaced higher in the ventricle with its tip reaching to the annulus. In both PMV and PLAMV, the chordae are short and thickened, thus restricting the motion of the leaflets.

arcade valve

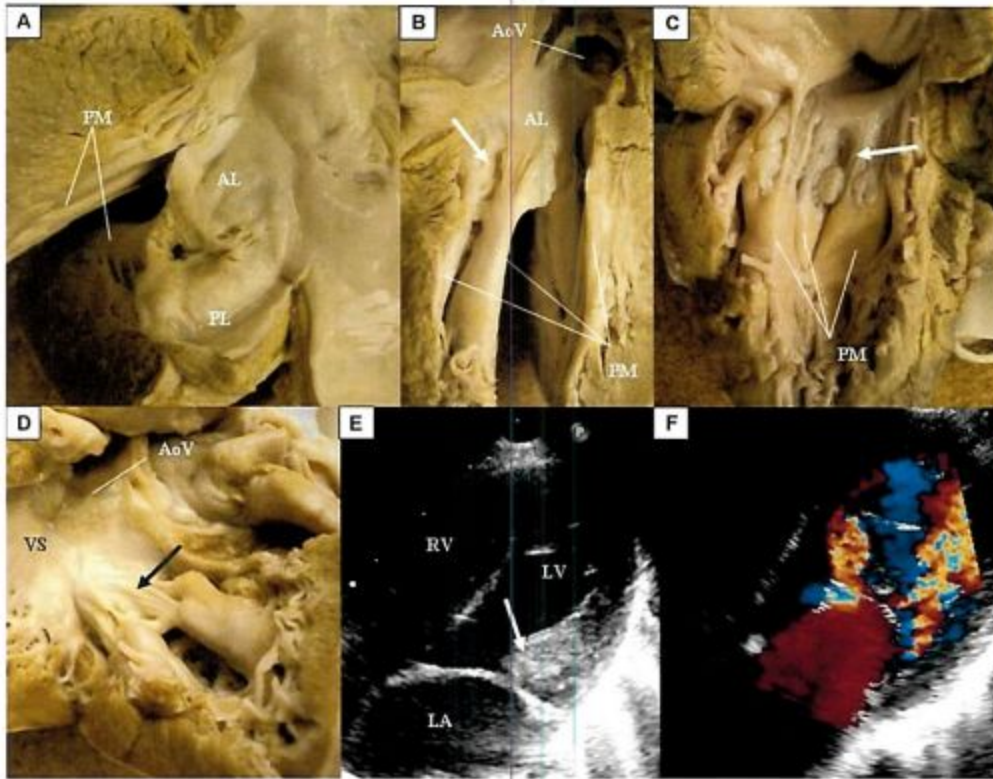


Figure 9. Arcade/hammock mitral valve. (A) Photograph showing the typical aspect of a hammock mitral valve seen from the left atrium and (B) the same valve seen from the left ventricle. (C and D) Postmortem specimens of anomalous mitral arcade characterized by fused interchordal spaces (arrows). (E) Two-dimensional echocardiographic view showing the obliterated interchordal spaces (arrow). (F) The typical aspect in Doppler colour mode of multiple jets through the reduced interchordal spaces. AoV: aortic valve; AL: anterior leaflet; LA: left atrium; LV: left ventricle; PL: posterior leaflet; PM: papillary muscles; RV: right ventricle; VS: ventricular septum.

coarctation of aov, valvular isovalvular aortic stenosis, mitral stenosis

Oosthoek et al. [13] assumed that PMV results from an embryological disturbance during the normal delamination of the trabecular ridge between the fifth and nineteenth week of gestation. In this hypothesis, the embryonic predecessors of the normal papillary muscles, derived from the anterior and posterior parts of the trabecular ridge, would condense into a single muscle. Although the spectrum of associated lesions is broad, PMV or PLAMV are commonly seen in association with other obstructive lesions affecting the left heart [56] or conotruncal anomalies [55]. As a consequence, the mitral valve should always be carefully inspected in order to diagnose PMV if any other feature of Shone's syndrome is present. Because opening of the mitral valve is limited, true PMV is highly associated with mitral stenosis. Mitral regurgitation occurs less commonly but must be equally carefully followed because of its progressive evolution. Because PMV is rarely diagnosed in isolation [55,56], asymptomatic cases are probably under-represented. Echocardiography establishes the diagnosis in

most patients with PMV. In the parasternal short-axis view, a single papillary muscle is confirmed at the mid-level of the left ventricle. The pathognomonic 'pear' shape of the mitral valve is seen in the four-chamber view, with the left atrium forming the larger base of the pear and the mitral leaflets the apex (Fig. 11) [57]. In this view, the valve has a typical 'domed' appearance in diastole. The majority (80%) of patients with PMV or PLAMV may not require surgical intervention in their first 10 years of life [56]. Conservative surgical treatment may consist of either chordal fenestration or papillary muscle splitting, associated or not with a commissurotomy [58]. When valvotomy is performed, the outcome closely depends on the size of the left ventricle. Indeed, left ventricular hypoplasia, classically described in cases of Shone's syndrome, has been proven to be a risk factor for poor outcome. Finally, true PMV is more correlated with univentricular palliation than PLAMV, because it is more often associated with left ventricular hypoplasia [55].

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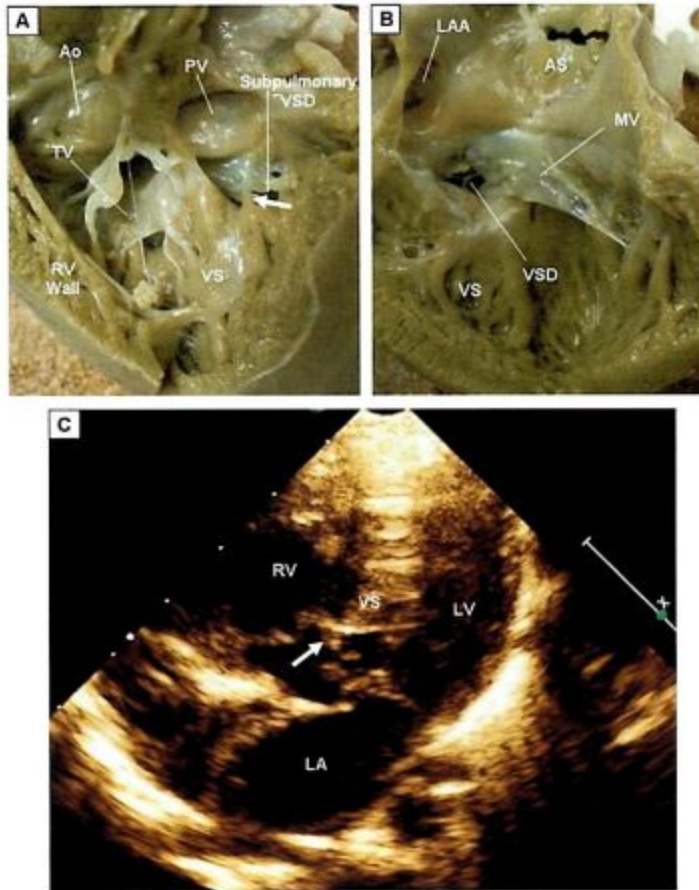


Figure 10. Straddling mitral valve. (A) Photograph of a straddling mitral valve associated with a double outlet right ventricle, seen from the right ventricle. The mitral valve is attached to the right ventricle by chordae (white arrow) that pass through the ventricular septal defect. (B) The same mitral valve seen from the left ventricle. (C) Echocardiographic view showing the abnormal attachment (white arrow) of the mitral valve in the right ventricle. Ao: aorta; AS: atrial septum; LA: left atrium; LAA: left atrial appendage; LV: left ventricle; MV: mitral valve; PV: pulmonary valve; RV: right ventricle; TV: tricuspid valve; VS: ventricular septum; VSD: ventricular septal defect.

Focus on the management of congenital mitral stenosis

The mitral valve is most commonly incompetent in all of these congenital anomalies. Mitral regurgitation is reported in 72% of cases, mitral stenosis in 13% and both stenosis and regurgitation in 15% [59]. Interventional therapies for medically refractory congenital mitral disease include percutaneous valvuloplasty, surgical valvuloplasty and mitral valve replacement. An intervention before the first year of life is rarely needed in cases of isolated regurgitation. In contrast, mitral stenosis may require early surgery. Age less than 1 year, hammock mitral valve and associated

cardiac anomalies are reported to be strong predictors of poor outcome [60]. Indeed, congenital mitral stenosis is rarely isolated [61] and is often associated with other left heart obstructions, thus being a part of a Shone's syndrome, in which the long-term surgical outcome is correlated with the severity of the mitral valve disease [62,63]. In his mitral ring series, Toscano et al. always found Shone's syndrome in cases of intramitral ring [45]. Finally, different congenital malformations of the leaflets, chordae tendinae and papillary muscles may be associated, making any procedure extremely difficult, especially in the newborn.

In children, mitral repair is always preferable to mitral valve replacement, even if the outcomes of this alternative seem to be acceptable [5,6]. Indeed, late outcomes of

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less than 1 year
↳ mitral stenosis
hammock mitral valve

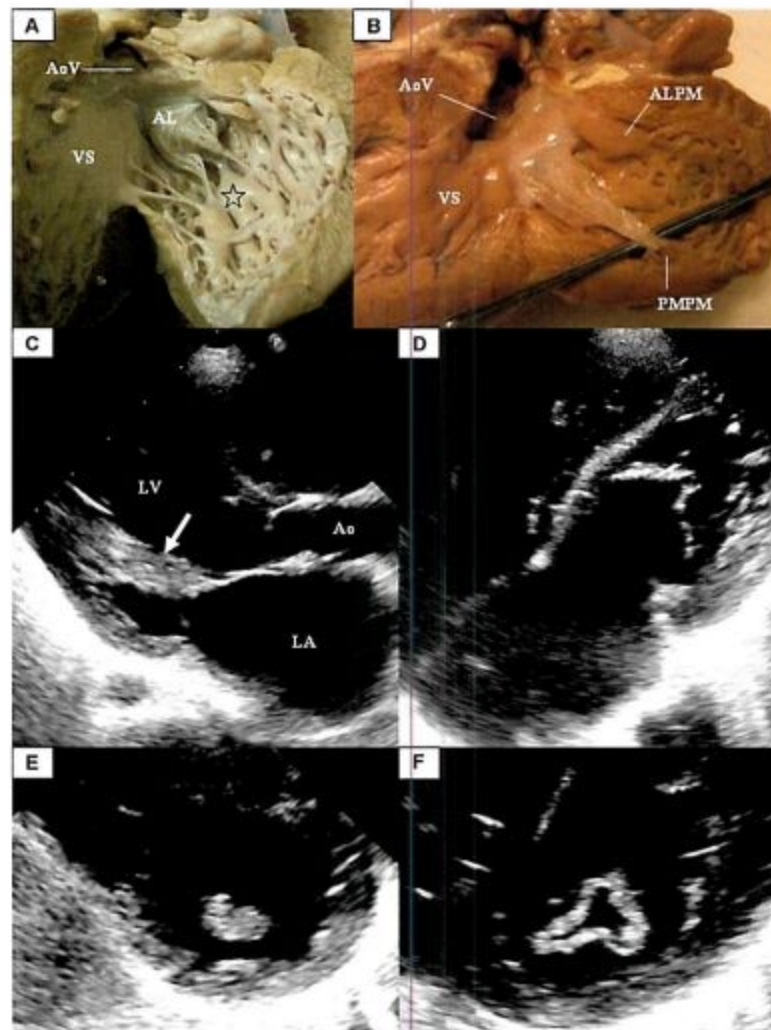


Figure 11. Parachute mitral valve. (A) Postmortem specimen of a true parachute mitral valve showing a fused papillary muscle (star). All chordae are inserted into this single papillary muscle. (B) Photograph of a parachute-like asymmetric mitral valve. The posteromedial papillary muscle is clearly underdeveloped. (C) Two-dimensional echocardiographic parasternal long-axis view showing a single papillary muscle connected to the leaflets by short and thickened chordae (arrow). (D) Apical four-chamber view showing the pathognomonic pear-shaped mitral valve. (E and F) Parasternal short-axis views showing the typical aspects of both the single papillary muscle and the mitral valve. ALPM: anterolateral papillary muscle; Ao: aorta; AoV: aortic valve; LA: left atrium; LV: left ventricle; PMPM: posteromedial papillary muscle; VS: ventricular septum.

valve repair are superior to replacement for both isolated congenital mitral anomalies [4] and associated anomalies [64]. Furthermore, mechanical valves require anticoagulation therapy, which may be very difficult to manage in small children. Percutaneous dilation of congenital mitral stenosis allows a significant decrease of the mitral gradient [59,65],

with mortality slightly better than that of surgical repair. However, this technique is not curative and requires re-intervention in 61% of cases at 5 years. This method may sometimes be useful in severe neonatal forms with combined lesions of the mitral valve. Severe congenital mitral stenosis is a rare and challenging condition, the optimal treatment

NON surgical, balloon, catheterisation

mechanical valves → anticoagulation therapy

of which is still debated) the result of the intervention also depends on the skill of the operator.

Conclusion

Different congenital malformations may affect the mitral valve either in isolation or in association with other cardiac anomalies. Improvements in surgical techniques have made it possible to obtain good results when a mitral repair is required. Anatomical analysis is of particular importance both for surgical management and prognosis.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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