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The Pathological Anatomy of Surgically Reconstructable or Prosthetically Correctable Congenital Valvular Malformation of the Mitral Region

E.-W. Schwarze and A. Bernhard

Department of Pathology (Head: Prof. Dr. K. Lennert) and
Department of Cardiovascular Surgery (Head: Prof. Dr. A. Bernhard), University of Kiel

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Summary. The special pathology of reconstructable or only prosthetically correctable congenital malformations of the mitral valve is described on the basis of the following examples taken from our own operative and autopsy material of the last 5 years:

1. Congenital isolated mitral stenosis in female twins (7 month old infant and 33 month old child).
2. Congenital isolated mitral insufficiency in a $7^1/_2$ year old boy.
3. Combined mixed mitral valve malformations with a parachute valve-like mitral valve anomaly, combined with hypoplasia of the ascending and descending aortas, in a $6^1/_2$ year old girl.
4. Congenital mitral insufficiency with a parachute mitral value, combined with supravalvular aortic stenosis and multiple peripheral stenoses of the pulmonary arteries in a $13^1/_4$ year old boy.
5. Insufficiency of the mitrally inverted tricuspid value with so-called corrected transposition of the great vessels in a 6 year old boy and with Ebstein's anomaly in a $2^1/_2$ year old boy.
6. A second mitral ostium in the aortic mitral leaflet with a partial atrioventricular canal in a $6^3/_4$ year old girl with Ellis-van Creveld syndrome.
7. Bland-White-Garland syndrome with relative mitral insufficiency in a 5 month old and a 4 month old boy.

Despite the recurrence of similar and comparable findings, each of our cases of congenital or early acquired noninfectious mitral valve malformation was formally different. This was also true for the cases of congenital isolated mitral stenosis in twins. Therefore, surgical correction requires a unique procedure for each case. It is possible to reliably infer the degree of malfunction of the atrioventricular valve in a mitral position from the special pathology only by considering the clinical data. On the other hand, a detailed evaluation of congenital mitral valve malformations is possible only through direct inspection—either by the surgeon or through an autopsy—despite modern cardiodiagnostic methods. Typical secondary findings are also discussed—for instance, endocardial fibrosis of the left atrium and the configuration of the heart. The anatomical prerequisites for surgical reconstruction or replacement of the valve with a prosthesis are mentioned.

Key words: Congenital or Early Acquired Noninfectious Mitral Valve Malformation — Congenital Isolated Mitral Stenosis — Congenital Isolated Mitral Insufficiency — Combined Mixed Mitral Valve Malformation — Insufficiency of Mitrally Inverted Tricuspid Valve with Ebstein's Anomaly — Second Mitral Ostium — Atrio-Ventricular Canal — Bland-White-Garland Syndrome — Endocardial Fibrosis — Heart Configuration — Pathological Anatomy.

Zusammenfassung. An folgenden Beispielen aus dem eigenen Operations- und autoptischen Untersuchungsgut der letzten 5 Jahre wird die spezielle Pathologie rekonstruierbarer oder prothetisch korrigierbarer kongenitaler Herzfehler der Mitralregion dargestellt:

1. Kongenitale isolierte Mitralstenose bei einem weiblichen Zwillingspaar (7 Monate alt gewordener Säugling bzw. $2^9/_{12}$ Jahre alt gewordenes Kleinkind),
2. kongenitale isolierte Mitralinsuffizienz bei einem $7^1/_2$jährigen Jungen,

3. kombiniertes, unreines Mitralvitium bei parachute-valveähnlicher Mitralklappenanomalie, kombiniert mit einer Hypoplasie der Aorta ascendens et descendens, bei einem $6^1/_2$jährigen Mädchen,

4. kongenitale Mitralinsuffizienz bei parachute mitral valve, kombiniert mit supravalvulärer Aortenstenose und multiplen peripheren Pulmonalarterienstenosen, bei einem $13^1/_4$ Jahre alten Jungen,

5. Insuffizienz der in Mitralposition invertierten Tricuspidalklappe bei sog. korrigierter Transposition der großen Gefäße bei einem 6 Jahre alt gewordenen Jungen und mit Ebsteinscher Anomalie bei einem $2^1/_2$jährigen Jungen,

6. zweites Mitralostium im aortalen Mitralsegel bei partiellem AV-Kanal bei einem $6^3/_4$ Jahre alt gewordenen Mädchen mit Ellis-van Creveld-Syndrom,

7. Bland-White-Garland-Syndrom mit relativer Mitralklappeninsuffizienz bei einem 5 Monate und bei einem 4 Monate alten männlichen Säugling.

Trotz der Wiederkehr ähnlicher oder vergleichbarer Befunde ist jedes der von uns beobachteten *kongenitalen oder frühkindlich* erworbenen, *nicht entzündlichen Mitralvitien* für sich gestaltlich verschieden. Die operative Korrektur erforderte daher in jedem Falle ein individuelles Vorgehen. Einerseits ist nur unter Verwendung der klinischen Untersuchungsbefunde aus der speziellen Pathologie verläßlich auf den Grad der Fehlfunktion der AV-Klappe in Mitralposition rückzuschließen. Andererseits läßt sich der detaillierte pathologisch-anatomische Befund bei kongenitalen Mitralvitien trotz der heute verfügbaren kardiodiagnostischen Methoden erst bei der direkten Inspektion — sei es unter der Sicht des Operateurs, sei es autoptisch — erheben. Neben dem jeweiligen Klappenbefund werden die für Mitralvitien typischen Sekundärveränderungen besprochen — so die links atrial betonte Endocardfibroelastose und die Herzkonfiguration. Auf die anatomischen Voraussetzungen für ein chirurgisch rekonstruktives Vorgehen oder einen prothetischen Herzklappenersatz wird hingewiesen.

Using the methods available today it is possible to clinically diagnose most of the congenital malformations of the heart that are compatible with a longer survival of the patients and therefore can be corrected surgically in some cases. However, a detailed anatomical description of congenital malformations of the mitral valve is often possible only through direct intraoperative or post-mortem inspection of the valvular apparatus. This is due to the fact that, despite the possible similarity of the clinical symptoms, every congenital mitral valve malformation has its own particular morphological features. In most cases it differs significantly from acquired mitral valve disease (Davachi *et al.*, 1971; Kilman *et al.*, 1971). However, in some cases, such as mitral stenosis, the distinction between a congenital and an acquired anomaly may be difficult (Doerr, 1960; Edwards *et al.*, 1965) Accordingly, the operative procedure has to be adjusted to fit the requirements of each case. This will be demonstrated here using examples from our own operative and autopsy material of the last 5 years.

Material

A. Congenital Isolated Mitral Stenosis in Twins

1. Patient A.K.R., Autopsy-No. 607/71

The 7 month old female infant (Twin I; 72 cm large, 5.9 kg in weight) with congenital isolated mitral stenosis had to undergo surgery because of cardiac failure that did not respond to conservative measures.

Autopsy Findings. Condition after recent valvotomy with a 0.9 cm long, longitudinal, paracommissural incision in the posterior mitral leaflet (Fig. 1). The mitral leaflet was thickened and bulgy and showed partly nodose and chondroid hardening near the mural insertion of the leaflet. The anulus fibrosus was not detectably hardened. Only the right commissure

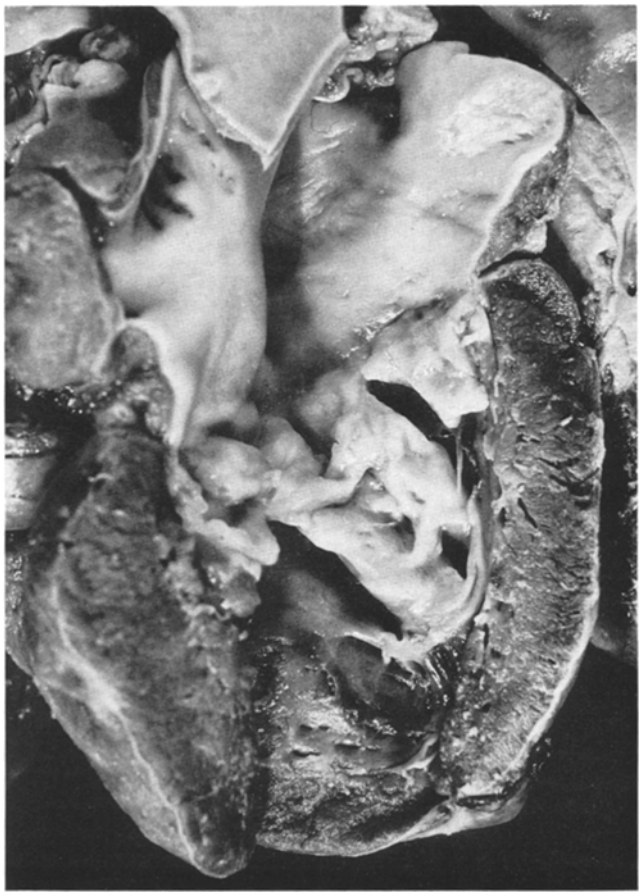


Fig. 1. Congenital isolated mitral stenosis. Condition after paracommissural valvotomy the day before death. The endocardium is thickened in the form of curtain folds, covers the chordae tendineae, and fuses these with each other hand the free margins of the valve with the papillary muscles together. Plump posterior papillary muscles are fused together through thickened endocardium. Diffuse endocardial fibrosis of the left atrium.—A.K.R., female, 7 months old, Twin I. Case 1

could be seen. The free margins of the valve were turned in. The endocardium was thickened in the form of curtain folds, covered the chordae tendineae, and fused these with each other and the free margins of the valve with the papillary muscles together. Near the apex two ascending, pillar-like, plump posterior papillary muscles of posterolateral origin were fused together through thickened cross-folded endocardium. There was a moderately thick, short anterior papillary muscle. The inner valvular circumference at the level of the free margins of the leaflet was 3.7 cm. The left atrium was clearly enlarged (left atrioventricular relation 1.5:1), showed thickening of the muscular wall (5 mm) and diffuse thickening of the endocardium to 1.5 mm (histologically: bland endocardial fibroelastosis). For further details cf. Schwarze (1975).

2. Patient M.R., Autopsy-No. 783/73

The 33 month old female child (Twin II; 98 cm large, 12.7 kg in weight) was operated on $6^1/_2$ months before her death because of congenital mitral stenosis.

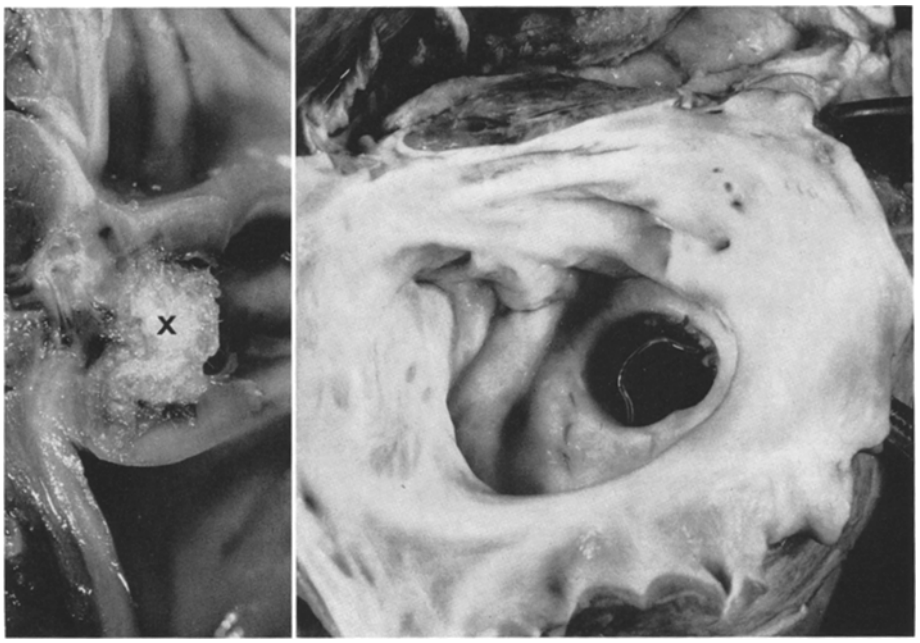


Fig. 2. Condition after total extirpation of the mitral valve apparatus because of congenital mitral stenosis and after implantation of a Björk-Shiley prosthesis $6^1/_2$ months before death. Verrucous parietal thrombi in the bed of the prosthesis (on the right, at 12 to 3 o'clock at the prosthesis ring). Endocardial bulge around the prosthesis ring, sagittal section (on the left), after removal of the metal prosthesis; synthetic ring (x) in situ.—M.R., female, 33 months old, Twin II. Case 2

Autopsy Findings. Condition after total extirpation of the mitral valve apparatus and after implantation of a Björk-Shiley prosthesis (17 MBRP). The edge of the prosthesis was bulgy and circularly overgrown with a fibrous, up to 3 mm thick endocardium which hindered the free movement of the prosthesis disc (Fig. 2). After removing the metal ring of the prosthesis one could see circular comb-like, organized thrombi in the bed of the prosthesis, some of which overlooked the endocardial bulge on both the atrial side (Fig. 2) and the ventricular side. The left atrium was large and spherical, the left ventricule was also large (inflow tract 4 cm long, outflow tract 5 cm long). There was almost diffuse thickening of the parietal ventricular endocardium with levelling of the trabecular surface especially underneath the mitral valve prosthesis; there was also indication of endocardial fibrosis in the left ventricular outflow tract. The left atrium was enormous and showed thickening of the muscular wall (6–7 mm) and diffuse endocardial fibrosis (1.0 mm). For further details cf. Schwarze (1975).

A histological investigation of a mitral valve biopsy $6^1/_2$ months before death of the patient showed no signs of florid infection.

B. Congenital Isolated Mitral Insufficiency

3. Patient Th.Th., Biopsy-No. 19 115/70

The $7^1/_2$ year old boy underwent a vital operation because of highly severe insufficiency of the heart due to congenital mitral insufficiency. Earlier, the valve had been replaced by a fascia lata according to the procedure of Ionescu and Ross. Three years later the fascia lata had to be replaced by a Björk-Shiley prosthesis (25 MBRP) due to endocarditis.

Intraoperative Findings. The left atrium was large and projected wide to the right, forcing the right atrium and the right ventricle away to the upper left. Mitral valve: There was a

cleft above the posterior commissure. The aortic leaflet was small, shrunken, and thickened. The mural leaflet was not transparent, rolled up, and stiff. There were no groups of papillary muscles, but instead 4–5 large parietal muscle projections, which sent out chordae tendineae to the mural and aortic leaflets.

C. Combined, Mixed Congenital Mitral Valve Malformation and Hypoplasia of the Ascending and Descending Aortas

4. Patient S.N.

The $6^1/_2$ year old girl underwent surgery because of cardiac insufficiency that could not be conservatively controlled due to a combined mitral valve malformation. The valve was replaced with an autologous fascia lata.

Intraoperative Findings. The heart was greatly enlarged with extreme enlargement of the left atrium. The right atrium and the right ventricle were also enlarged. The ascending and descending aortas were hypoplastic. Mitral valve: The tissue was delicate, both leaflets were transparent. Chordae tendineae of both leaflets inserted at one very large papillary muscle which originated from the region around the apex of the heart. A second papillary muscle could not be definitely recognized, instead there was a trabecula with insertion of chordae tendineae of the mural and aortic leaflets. There was a cleft in the aortic leaflet near the posterior commissure.

D. Congenital Mitral Insufficiency, Supravalvular Aortic Stenosis, and Multiple Peripheral Pulmonary Arteriostenoses

5. Patient J.H.

The $13^1/_4$ year old boy had to undergo surgery because of progressive dyspnea under stress.

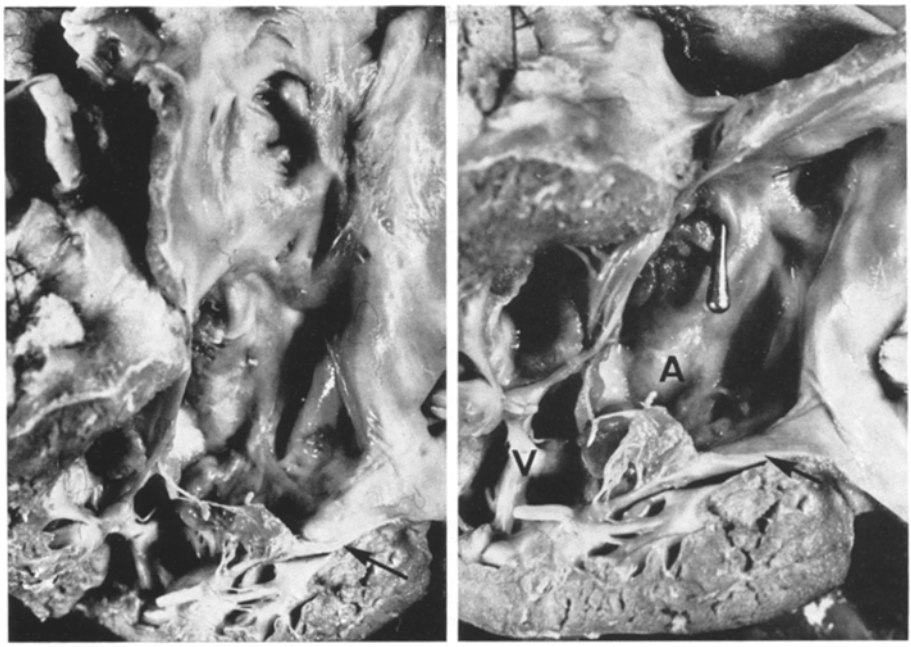
Intraoperative Findings. The left ventricle was enlarged, projected wide to the left, and showed severe muscular hypertrophy. The bulbus of the aorta showed a waist. Mitral valve: The aortic leaflet was thickened and club-shaped, the mural leaflet was also thickened. Deep in the left ventricle, near the apex of the heart, there was an approximately 1 cm thick, singular papillary muscle, at which all of the chordae tendineae were inserted. The mitral leaflets were drawn into the left ventricle in the shape of a funnel (so-called parachute mitral valve). The mitral valve had to be removed and a Björk-Shiley prosthesis was implanted.

E. Corrected Transposition of the Great Vessels with Insufficiency of the Mitrally Inverted Tricuspid Valve in Ebstein's Anomaly

6. Patient W.Z., Autopsy-No. 53/72

The $2^1/_2$ year old boy had a corrected transposition of the great vessels. He underwent surgery because of an accompanying ventricular septal defect (50% shunt volume).

Autopsy Findings. The whole heart was enlarged. The atria were in normal position. Pulmonary veins and venae cavae entered typically. The ventricles were inverted: the functionally right, anatomically left ventricle was slightly displaced to the rear and had a bicuspid atrioventricular valve in reversed position. The functionally left, anatomically right ventricle was slightly displaced to the front and had a tricuspid atrioventricular valve (reversed tricuspid valve) with Ebstein's anomaly: the plane of insertion of this valve was displaced deep into the functionally left ventricle (Figs. 3 and 4). Compared to the septal and anterior tricuspid leaflet, the dorsal tricuspid leaflet was deeply inserted with very short chordae tendineae and was highly hypoplastic. There was ectasia of the fibrous ring of the reversed tricuspid valve with balloonlike widening of the supravalvular "atrialized" left ventricular inflow tract. The left atrium showed severe dilatation and hypertrophy. There had been recent subtotal surgical occlusion of an abnormally high ventricular septal defect in the conus arteriosus pulmonalis (Fig. 4, probe!). There was severe ectasia of the pulmonary trunk, the pulmonary arteries, and of their branches. The great vessels were transposed and inverted. Coronary arteries with a



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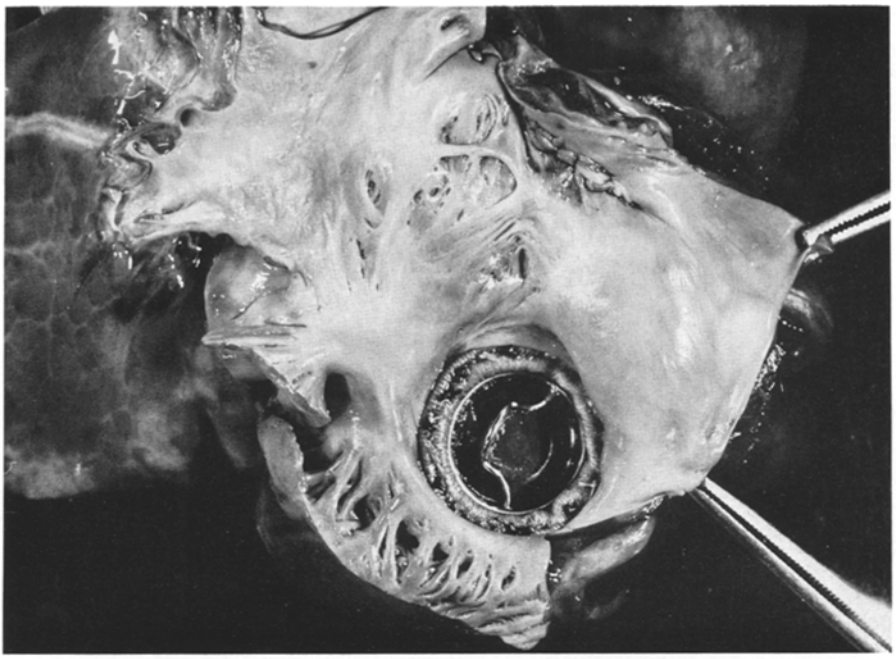
Figs. 3 and 4. Ebstein's anomaly of the mitrally inverted tricuspid valve with corrected transposition of the great vessels. The valve and the plane of insertion (arrow!) are displaced deeply into the functionally left, anatomically right ventricle. VSD (probe!) with a right ventricular-left atrial shunt. "Atrialized" supravalvular and valvular inflow tract (*A*) of the left ventricle (*V*).—W.Z., male, $2^1/_2$ years old. Case 6

topographically inverted course arose in reversed position from the dorsal aortic sinus of Valsalva. The foramen ovale was closed, the ductus arteriosus obliterated.

7. Patient K.Sch., Autopsy-No. 642/74

The 6 year old boy with a corrected transposition of the great vessels underwent surgery due to severe tricuspid valve insufficiency.

Autopsy Findings. The heart was enlarged, particularly on the left side. The superior and inferior venae cavae entered a relatively small, anatomically right atrium, which was located in a right dorsal position. The pulmonary veins entered the greatly enlarged left atrium, which lay in a ventrolateral position. The ventricles were inverted: the right, anatomically left ventricle was oblique, relatively narrow, muscular, and in dorsal position. The right atrioventricular valve appeared as a reversed and dysplastic mitral valve; its anterior leaflet continued into the right and posterior semilunar cusp of the pulmonary orifice. The patient had undergone infundibulectomy because of arch-like, fibrous and muscular subvalvular pulmonary stenosis. The left, anatomically right ventricle was in a ventrolateral position and was highly dilated. The left atrioventricular valve had been recently removed and replaced with a Björk-Shiley prosthesis (Figs. 5 and 6). The left atrium showed severe ectasia and moderately severe diffuse endocardial fibrosis (Fig. 5). The great vessels were transposed and inverted. There was monostial origin of the coronary arteries from the right sinus of Valsalva; they showed an atypical distribution. The foramen ovale was closed, the ductus arteriosus obliterated.



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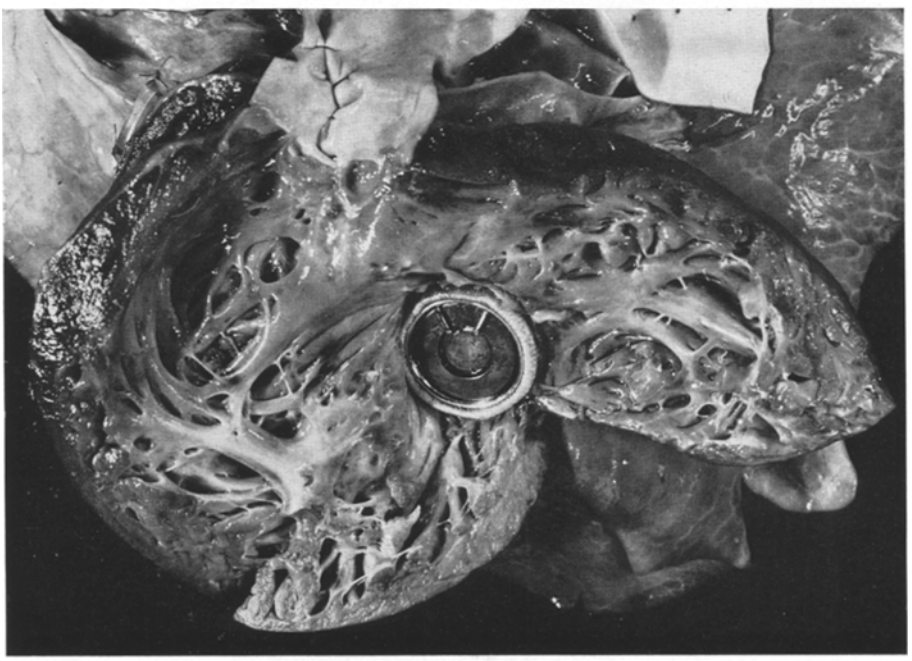

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Figs. 5 and 6. Condition after total extirpation of the tricuspid atrioventricular valve apparatus because of valve insufficiency in a case of corrected transposition of the great vessels. Implantation of a Björk-Shiley prosthesis. Death in tabula. Diffuse endocardial fibrosis of the left atrium, arterialization of the pulmonary veins. Hypertrophy, dilatation, and local endocardial fibrosis of the functionally left, anatomically right ventricle.—K.Sch., male, 6 years old. Case 7

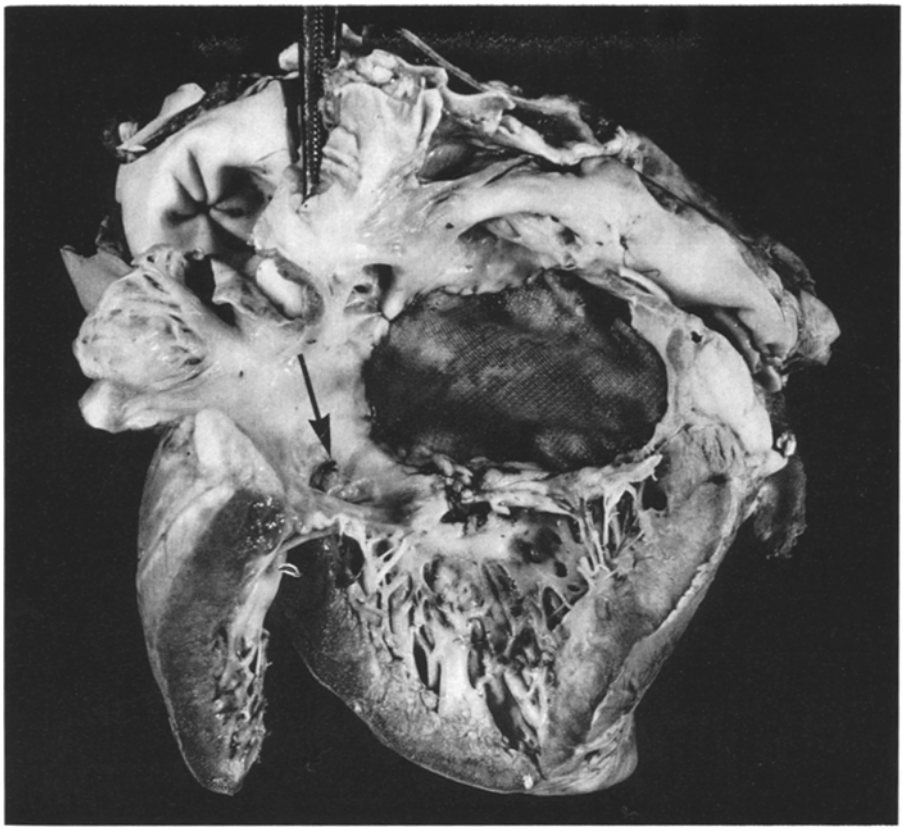
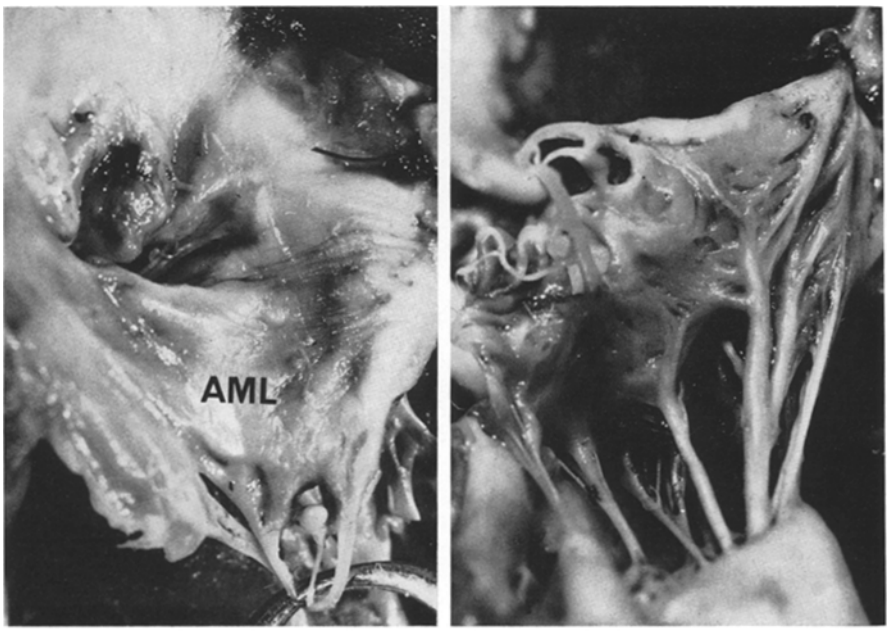


Fig. 7. Left atrium and ventricle. Condition after closure of a partial atrioventricular canal with a patch 3 days before death. A second mitral ostium (arrow!) in the anterior mitral leaflet.—M.T., female, $6^{3}/_{4}$ years old. Case 8



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F. Second Mitral Ostium in the Mitral Valve in a Case of Endocardial Cushion Defect (Here: Partial Atrioventricular Canal)

8. Patient M.T., Autopsy-No. 574/73

The $6^3/_4$ year old girl showed signs of chondroectodermal dysplasia (Ellis-van Creveld syndrome). She underwent surgery 3 days before death because of pulmonary hypertension due to an atrial septal defect and mitral insufficiency.

Autopsy Findings. The heart was on the whole slightly enlarged with a spread waist and a wide right and left atrium. The right ventricle was wide and turned toward the front. The atrial septum was completely missing. The partial atrioventricular canal had been closed with a patch (Fig. 7). Mitral valve: The aortic and mural leaflets were attached to the instep of the ventricular septum. The transition between the mural leaflet and the posterior tricuspid leaflet was almost continuous. Short chordae tendineae ran from the parts of both mitral leaflets near the posterior commissure to the part of the ventricular septum near the instep. The free margins of the mitral leaflet were rolled up. In the aortic mitral leaflet there was a second mitral ostium, 1 cm in diameter (Fig. 7, arrow!), with circular insertion of delicate chordae tendineae which ran to the anterior papillary muscle (Fig. 8). The left ventricular inflow tract was shortened, the outflow tract was of normal length but narrowed. Tricuspid valve: The septal leaflet was rudimentary, the other leaflets were normal. The right ventricle was wide and muscular with a flattened trabecular surface and an oblique outflow tract ascending to the left front. The foramen ovale was closed, the ductus arteriosus obliterated.

G. Relative Mitral Insufficiency in Bland-White-Garland Syndrome (Origin of the Left Coronary Artery from the Pulmonary Trunk)

9. Patient G.K., Autopsy-No. 915/70

The 5 month old male infant (67 cm large, 6.5 kg in weight) underwent thoracotomy for the purpose of ligating an atypical left coronary artery which arose from the pulmonary trunk with a transcoronary aortopulmonary shunt. The patient died during surgery.

Autopsy Findings. The heart was much larger in size than the child's fist. The left atrium and the left ventricle which formed the apex were particularly enlarged. The ostia were normal in size with typically formed, delicate atrioventricular valves—however, angiographically there was a left ventricular/left atrial reflux. The left coronary artery showed an atypical origin from the sinus of the septal semilunar cusp of the pulmonary orifice and was drawn toward the left ventricle in the sulcus coronarius. The right coronary artery had a wide lumen and showed a typical origin. Angiographically it was divided into three sinuous branches, anastomosing with the left coronary artery. A large part of the parietal endocardium was thickened, particularly above the papillary muscles of the dilatated left ventricle. There were disseminated spotty myocardial scars in the left ventricular wall. The foramen ovale was open only a slit. The ductus arteriosus could be passed through with a small probe.

10. Patient S.D., Autopsy-No. 306/73

The 4 month old male infant (64 cm large, 4.5 kg in weight) underwent surgery 3 weeks before death for the purpose of ligating the coronary artery and correcting the mitral valve. In this case of Bland-White-Garland syndrome there was severe mitral insufficiency.

Intraoperative Findings. The left side of the heart was exaggerated with enlargement of the left and right ventricles and enormous dilatation of the left atrium, which projected in a dorsal and left lateral direction. There was a large calibered coronary arterial plexus between the right atrium and the right ventricle and on the latter itself. The left coronary artery was as thin as a knitting needle and arose dorsally above the level of the valve from the pulmonary trunk. Mitral valve: The valve ring was wide, the aortic leaflet transparent. The mural leaflet was nodulated and rolled downward. No real papillary muscle could be seen; instead the chordae tendineae of both leaflets seemed to arise directly from the mural trabeculae.—Surgical reconstruction of the mitral valve was impossible. A Björk-Shiley prosthesis (17 MBRP) was implanted.

Fig. 8a and b. Details of Fig. 7. (a) Lateral view from above: ostium and funnel of the second mitral ostium. Anterior mitral leaflet (*AML*). (b) Anterior view from below: thickened chordae tendineae pulled together to a common basis (parachute-like valve)

Autopsy Findings. The left coronary artery arose from the sinus of the left semilunar cusp of the pulmonary orifice. In the distribution of the coronary arteries there was predominance of the right one. The stump of the anterior papillary muscle was shrunken, scarred, and calcified. There were multiple disseminated myocardial scars in the whole left ventricular wall. The left ventricle showed moderately strong dilatation, the left atrium dilatation and diffuse endocardial fibrosis. The right ventricle was hypertrophic and dilatated. The foramen ovale was closed, the ductus arteriosus obliterated.

Discussion

Patho-anatomically, malfunction of the atrioventricular valve in cases of congenital mitral valve malformation is based on an anomaly in the form or position of some or all parts of the apparatus of the mitral valve. The atrioventricular valves, the chordae tendineae, and/or the papillary muscles can be anomalous. Despite the recurrence of similar and comparable findings, a synopsis of our cases shows that there is no set of uniform characteristics for congenital and early acquired noninfectious mitral valve malformations; instead, each case is different. This is also true for isolated congenital mitral stenosis in twins, which we recently described in detail (Schwarze, 1975). The cases of congenital mitral stenosis reported in the literature (Ferencz *et al.*, 1954; Daoud *et al.*, 1963; Edwards *et al.*, 1965; Prado *et al.*, 1965; Beuren, 1966; Diekmann *et al.*, 1969; Davachi *et al.*, 1971; Roberts and Perloff, 1972) often showed a hypoplastic apparatus of the mitral valve. In such cases the valvular tissue was thickened in bulges. It could also show nodose or chondroid hardening. In a few cases the leaflets were transparent and not deformed. The commissures were indistinct or not developed at all. The chordae tendineae were thickened, shortened, and often fused together. In our Case 1 they were hidden under endocardium thickened in the form of curtain folds (Fig. 1). They could also be completely absent, in which case the free margins of the leaflets were attached directly to the papillary muscles. The papillary muscles could be either delicate or plump. In the latter case—as in our Case 1—this causes subvalvular as well as valvular stenosis (Castaneda *et al.*, 1969; Davachi *et al.*, 1970). The number and position of the papillary muscles varied. They were sometimes absent. In these cases the chordae tendineae inserted separately at the leaflets and ectopically at the mural endo- or myocardium. On the other hand, there could also be numerous (sometimes 4 to 5 per leaflet) papillary muscles, occasionally reduced to trabecula-like ridges. Sometimes there was only one papillary muscle, often originating near the apex, at which all of the chordae tendineae were inserted. Various forms of congenital mitral stenosis develop in cases with a normally wide or narrowed valve ring (Edwards *et al.*, 1965): a diaphragm-, chimney-, or funnel-like stenosis (Starkey, 1959).

The congenital diaphragm-like stenosis can be similar to acquired rheumatic mitral stenosis (Edwards *et al.*, 1965). The so-called parachute mitral valve is a typical representative of funnel-like stenosis (Shone *et al.*, 1963). Here the chordae tendineae are inserted at a solitary papillary muscle arising near the apex and pull the usually delicate leaflets concentrically into the middle of the left ventricle (see Cases 4 and 5). When the chordae tendineae are short and also fused together, the passage of blood at the bottom of the funnel is hindered, the valve is functionally stenotic (Davachi *et al.*, 1971). The valve can also be insufficient (Cases 4

and 5)—depending on the form and insertion of the chordae tendineae—or functionally competent (Glancy *et al.*, 1971).

The classical case of a parachute mitral valve described by Shone *et al.* (1963) was combined with subvalvular ring stenosis of the left atrium, subaortic stenosis, and stenosis of the isthmus of the aorta. If not isolated, the parachute mitral valve also occurs with only one or two of the mentioned malformations or with other accompanying anomalies: We believe that our Case 4 represents the first to be described combination of a parachute mitral valve with supravalvular aortic stenosis and multiple peripheral pulmonary arteriostenoses. Supravalvular aortic stenosis is occasionally combined with malformations of the mitral valve (Becker *et al.*, 1972).

Congenital mitral insufficiency is more frequent than isolated congenital mitral stenosis and, like congenital mitral stenosis, more often combined with other cardiovascular anomalies than isolated. Anomalies often observed together with congenital mitral insufficiency are: atrial septal defect (ASD I), atrioventricular canal, corrected transposition of the great vessels, primary (idiopathic) endocardial fibrosis with involvement of the apparatus of the mitral valve, stenosis of the isthmus of the aorta, aortic stenosis, and patent ductus arteriosus (Beuren, 1966; Hilgenberg *et al.*, 1972; Buttler *et al.*, 1973).

Of the 8 cases of congenital or early acquired mitral valve malformation that we have observed in the last 5 years, mitral insufficiency was the most predominant with 4 cases (or 5 cases if one includes one case of combined mitral valve malformation) as against 2 cases of mitral stenosis (twin sisters with congenital isolated mitral stenosis). Of the 4 (or 5) cases with mitral insufficiency there was only one case of isolated congenital mitral insufficiency (Case 3).

As described for mitral stenosis, anomalies of the whole or part of the apparatus of the valve can cause the mitral insufficiency (Berghuis *et al.*, 1964; Schieken *et al.*, 1971). In addition to and typical for mitral insufficiency are further: partial or total cleft in the mitral leaflet (see Cases 3 and 4), double mitral ostium (Case 7; Figs. 5 and 6), longer than normal, lasso-like chordae tendineae and/or atypical origin of the chordae tendineae and leaflet (Case 8).

The left atrioventricular valve apparatus is often malformed when the tricuspid valve is ectopic and inverted in a mitral position. In our Case 6 of Ebstein's anomaly, for instance, there was an abnormally deep origin of the leaflet far below the anulus fibrosus (Figs. 3 and 4) and shortening and ectopic insertion of the chordae tendineae, which fixed the leaflet to the parietal endocardium. With narrowing of the left ventricle this leads to an irregularly shaped valvular infundibulum, which can simulate mitral insufficiency when it functionally belongs to the atrium, or which can also be connected with a real mitral insufficiency. The special anatomy of Ebstein's anomaly makes the replacement of the valve with a prosthesis problematical (Berry *et al.*, 1974).

Just as a hypoplastic left heart (not considered in our study) is connected with hypoplasia/stenosis, or in extreme cases with atresia of the mitral valve, endocardial cushion defects are as a rule paired with a disturbance in the development of the mitral valve (Mierop, 1962; Goerttler, 1963; Baron, 1972). This does not necessarily cause valvular functional incompetence (Frater, 1965; Perloff in Roberts and Perloff, 1972). However, in our case (Case 7) with a partial atrio-

ventricular canal the mitral insufficiency was caused by dysplasia of the mitral leaflet and a second ostium in the aortic leaflet (parachute valve-like second mitral valve in the mitral valve).

Bland-White-Garland syndrome is comparable to relative mitral insufficiency in adults after an infarct or myocarditis that has taken its course (scarred heart). It is characterized by anomalous origin of the left coronary artery from the pulmonary trunk (Cases 9 and 10), which leads to arterial ischemia of the part of the myocardium supplied by this coronary artery. The blood is venous and the circulation may be inadequate due to a direct aortopulmonary shunt in cases of anastomosis of both coronary arteries. This means that there can be different patho-anatomical findings in parallel cases such as those reported by Sokolowska-Pituchowa *et al.* (1962) and our two cases: widespread cicatrization and calcification of the left ventricular anterior wall including the anterior papillary muscle and/or disseminated scars of the whole left ventricular wall including both papillary muscles. Atrophy of the papillary muscle (Case 10) with binding of the mitral leaflet, increased through the dilatation and subinvolution due to cicatrization of the left ventricular wall (Case 10), causes relative mitral insufficiency (Burchell and Brown, 1962; Wesselhoeft *et al.*, 1968). Both of our cases of Bland-White-Garland syndrome showed mitral insufficiency. This indicates that in every case of mitral insufficiency which develops in early childhood one has to consider the possibility of this syndrome!

The endocardial fibrosis of the left ventricle often observed in cases of Bland-White-Garland syndrome supports for instance the assumption that idiopathic endocardial fibrosis is an expression of hypoxemic damage (Andersen and Kelly, 1956; Still, 1961; Remmele, 1962; Goerttler, 1963; Moller *et al.*, 1964).

According to the reports in the literature, both primary idiopathic endocardial fibrosis and secondary endocardial fibrosis can be observed in cases of mitral stenosis. Endocardial fibrosis of the left atrium has been found in every case of congenital mitral stenosis—as in our Cases 1 and 2 (see Fig. 2)—and is considered to be secondary (Ferencz *et al.*, 1954). Endocardial fibrosis of the left ventricle, on the other hand, is rare (Beuren, 1966), particularly when there is no concurrent aortic stenosis (Ferencz *et al.*, 1954). Ventricular endocardial fibrosis can be observed, as in our Case 2, when the mitral valve has been replaced with a prosthesis (cf. Roberts and Morrow, 1968). As long as the etiology of diffuse endocardial fibrosis of early childhood and its relation to other diseases have not been clarified—which they still have not been (Folger, 1971)—we shall have to follow Moller *et al.* (1964) and speak of endocardial fibrosis on a purely descriptive basis. In both of our cases of congenital isolated mitral stenosis (Cases 1 and 2) the thickened endocardium showed a typical sequence of the layers, but the proportion of the individual layers was quite different from the norm. The fibromuscular and muscular layers were highly developed.

In Case 2 death occurred six months after implantation of a Björk-Shiley prosthesis due to malfunction of the prosthesis caused by a circular fibrous endocardial bulge on both the atrial and ventricular side of the prosthesis (Fig. 2). The bulging endocardial fibrosis may have been favored by a local thrombosis (Fig. 2), which was found circularly around the bed of the prosthesis after it was removed. Since the child suffered from postoperative serum hepatitis, anticoagulants could not be applied on a regular basis in sufficient doses.

The configuration of the heart in cases of congenital mitral valve malformation is similar to that of acquired malformations. It is determined by the enlargement of the left atrium and the hypertrophy of the right ventricle. The atria were moderately enlarged in our cases of congenital isolated mitral stenosis. There was enormous enlargement of the atrium, seen otherwise only in acquired mitral valve malformations, in our Case 4 of combined mitral valve malformation with a parachute valve-like anomaly of the mitral valve. In cases of endocardial cushion defects, on the other hand, the abnormal hemodynamics give the heart its form, mostly due to the enlargement of the right atrium.

A synopsis of our patho-anatomical findings shows that congenital or early acquired malformations of the mitral valve in infancy can be surgically reconstructed only when there is enough flexible valve material, when the papillary muscles are approximately normal, and when the fibrous valve ring is not narrowed to any great extent. Otherwise, the valvular malformation or deformation must be corrected prosthetically, as in most of our cases (cf. Cases 2, 3, 4, 5, 7, and 10).

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Dr. med. E.-W. Schwarze
Pathologisches Institut der Universität
D-2300 Kiel
Hospitalstr. 42
Federal Republic of Germany