Supravalvar mitral ring with a parachute mitral valve and subcoarctation of the aorta in a child with hemodynamically significant VSD. A study of the morphology, echocardiographic diagnostics and surgical therapy

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Abstract

The authors present a case of echocardiographic diagnosis of supravalvar mitral ring (a fibromembranous structure that arose from the atrial surface of the mitral leaflets) in a child with a parachute mitral valve, a ventricular septal defect, and mild narrowing of the aortic isthmus. The supravalvar mitral stenosis is a typical but very infrequently detected element of the complex of anatomical abnormalities located within the left heart and the proximal aorta, called the Shone's complex (syndrome). Diagnosing an additional, hemodynamically significant anatomic defect during echocardiography was possible thanks to the detection of marked mobility limitation of the ring-adjacent part of the mitral valve mural leaflet as well as of an atypical image of turbulence occurring during the inflow from the left atrium to the left ventricle. The early diagnosis made it possible to perform complete correction of this complex congenital defect within a single operation.

Congenital defects of the mitral valve hindering the inflow from the left atrium to the left ventricle constitute only 0.6% of all the congenital heart defects[1]. Clinical reports describing the supravalvar mitral ring as a specific morphological form of mitral stenosis are rare[2].

Case description

An eighteen-month-old girl was referred to the Department of Cardiac Pediatric Surgery, Medical University of Warsaw for the purpose of a surgical correction of a hemodynamically significant ventricular septal defect (VSD) coexisting with a parachute mitral valve with a slight degree of narrowing. In the preliminary echocardiographic examination it was assumed that the reason for the acceleration of blood inflow from the left atrium to the left ventricle was above all the increased volume of pulmonary venous return caused by VSD.

The transthoracic echocardiographic examination which is the principal examination qualifying for surgery, con-

firmed the presence of a big, infracristal defect in the interventricular septum (Fig. 1) and of a parachute mitral valve opening over the posteromedial papillary muscle without the visible residual muscle nor the anterolateral commissure (Fig. 2 and Fig. 3). Moreover, the examination demonstrated elongated tendinous cords which hindered the interpretation of the anatomic details of the valve.

The continuous-wave Doppler was used to demonstrate significant gradient through the mitral valve: maximum – 14 mm Hg, mean – 9 mm Hg. The attention of the examining _echocardiographer was drawn by the fact that the turbulence and the acceleration of inflow into the left ventricle appeared just above the level of the mitral valve ring and not at the level of the free edges of the mitral leaflets as one should expect if the narrowing was caused by the insufficient separation of leaflets. A more thorough analysis (the assessment of the image displayed in slow motion and "frame by frame") revealed the presence of a membranous, 2–4 mm high structure growing into the atrial lumen and connected with the left-atrial surface of the mitral leaflets
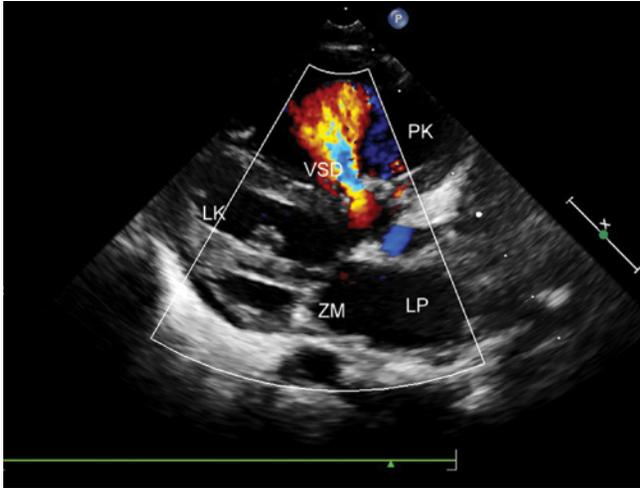
Fig. 1. *Parasternal long axis view of the left ventricle during the systole. Visible subaortic defect in the interventricular septum with a turbulent left-to-right shunt with some degree of aortic dextroposition. Closed leaflets of the mitral valve do not show obvious abnormalities. The presence of elongated tendinous cords connected with the subvalvular mitral apparatus. LP – left atrium, LK – left ventricle, ZM – mitral valve, PK – right ventricle, VSD –ventricular septal defect*
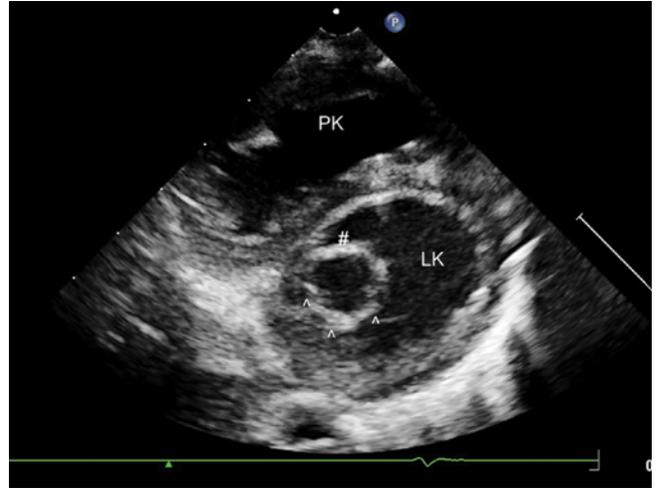
Fig. 2. *Parasternal transverse view at the level of the mitral valve leaflets. Visible open, nearly circular valve orifice located eccentrically – above the posteromedial papillary muscle. It is not possible to differentiate the commissures. The cross-section at this level does not show abnormal structures connected with the valve leaflets. LK – left ventricle, PK – right ventricle, # – aortic leaflet, ^ ^ – mural leaflet*

(Fig. 4). This membrane caused the narrowing of the mitral inflow canal above all as a result of the limitation of the diastolic mobility of the mitral leaflets – in the highest degree – of the mural (posterior) leaflet close to its base (Fig. 5, Fig. 6, Fig. 7). The pathological structure ran along the ring fragment located above the posterior-inferior wall (Fig. 8) and affected only a small part of the anterior (aortic) leaflet, impairing its function to a much lesser extent. Within the scope of the mural leaflet it caused a clear division into two parts: the one which was almost motionless, located between the valvar ring and the membrane and the free part of the leaflet maintaining correct mobility (Fig. 5, Fig. 6, Fig. 7). The precise assessment of the spatial relationship between the membrane and the leaflets was hindered by the abnormal structure of the parachute mitral valve which did not show signs of the presence of clear commissures which made it impossible to make a clear demarcation between the leaflets. In the apical four chamber view the obtained image suggested that the membrane joined both mitral leaflets whereas the view showing the long axis of the left ventricle and transverse views demonstrated this membrane's connection only with the mural leaflet. Additionally attention was drawn to the elongation of the distal part of the transverse aortic arch and to the borderline diameter of the aortic isthmus (Fig. 9) with the acceleration of the flow to the value of 1.95 m/s, with the maintaining of exclusively the systolic, correct flow spectrum in the abdominal aorta. The supravalvar narrowing of the parachute mitral valve coexisting with the abnormal anatomy of the aortic arch and aortic isthmus allowed for determining that the defect belongs to the spectrum of Shone's syndrome.

The child was qualified for VSD correction, the resection of the supravalvar mitral ring and the intraoperative assessment of the mitral valve anatomy in order to determine the

necessity of the potential separation of the single papillary muscle. Intraoperatively the presence of an oval intracristal defect in the interventricular septum was found, the dimensions of which were 4 × 10 mm; it was closed with a PTFE patch applying transatrial approach. The transeptal approach through the interatrial septum allowed for obtaining full visibility of the left atrium and especially the fibrous tissue ring the height of which was ca. 4 mm and which included nearly 2/3 of the parachute mitral valve perimeter. The performed resection of the ring resulted in obtaining satisfactory widening of the mitral orifice. No intervention was performed in the subvalvular apparatus.

Fig. 3. *Parasternal transverse view at the level of a single posteromedial papillary muscle. No structures connected to the anterolateral muscle have been revealed. LK – left ventricle, PK – right ventricle, MP – papillary muscle*
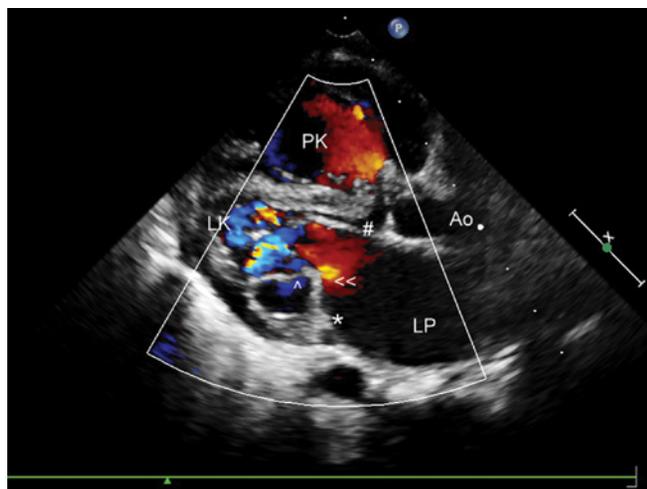
Fig. 4. *Parasternal long axis view of the left ventricle – diastole. Flow coded with color. Visible incomplete opening of the mural leaflet of the mitral valve and turbulence of the inflow to the left ventricle from the level of the excrescence on the atrial surface of this leaflet marked with the symbol „<<". A part of the leaflet between the ring and the excrescence practically does not show movement during diastole. The remaining part of the leaflet opens freely. LP – left atrium, LK – left ventricle, PK – right ventricle, Ao – aorta, * – the mitral valve ring, << – excrescence on the mural leaflet of the mitral valve, ^ – the movable part of the mural leaflet, # – the aortic mitral leaflet*

Fig. 5. *Parasternal view demonstrating the left atrium in the long axis – diastole. Visible incomplete separation of the mural leaflet of the mitral valve from the level of the excrescence on the atrial surface of this leaflet marked with the symbol „<<". A part of the leaflet between the ring and the excrescence practically does not show movement during diastole. The remaining part of the leaflet opens freely. Imaging without the color Doppler shows larger distance between the edges of the mitral valve leaflets than between their central parts due to the limitation of the mobility by the pathological membrane. LP – left atrium, LK – left ventricle, PK – right ventricle, Ao – aorta, * – the mitral valve ring, << – the excrescence on the mural leaflet of the mitral valve, ^ the movable part of the mural leaflet, # – the aortic mitral leaflet*

Postoperative course – without complications. Examinations performed after the correction demonstrated marked improvement of the mobility of the mitral valve leaflets and the lowering of the maximum gradient to 9 mm Hg and the mean one to 5 mm Hg (Fig. 10, Fig. 11). The child remains under outpatient observation of the Pediatric Cardiology Department of the Medical University of Warsaw.

Discussion

In the presented case the pathology of the mitral valve was composed of two elements: the parachute valve as well as the fibrous ring connected with the left-atrial surface of the mitral leaflets. Congenital mitral stenosis is a rare defect – it has been described in ca. 0.6% of autopsies of patients with congenital heart defects and 0.21–0.42% of clinical reports[1]. On the basis of 49 autopsy studies Ruckman and VanPraagh distinguished 4 types of congenital mitral stenosis[1,3]:

1) the „typical" form – characterized by the shortening of the tendinous cords, the obliteration of the space between the cords and the reduction of the distance between papillary muscles;
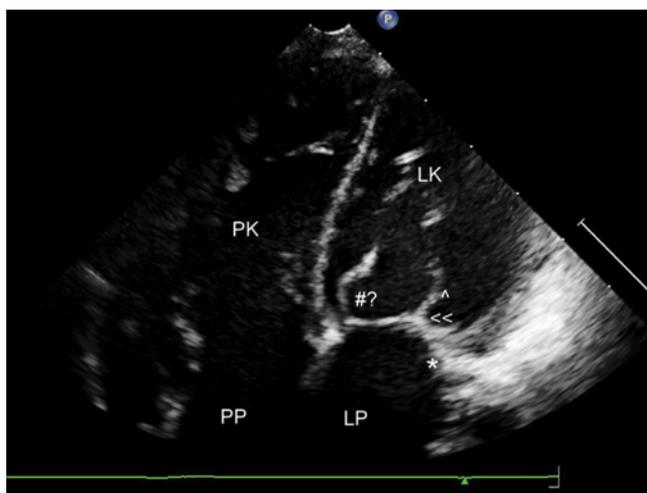
Fig. 6. *The apical four chamber view. Visible membranous structure connecting the bases of both leaflets of the mitral valve. Images obtained in parasternal views presenting the long axis of the left ventricle and the transverse ones generate doubts whether indeed this view demonstrates two different leaflets or whether a cross-section is obtained through different parts of the same mural mitral leaflet. This is the reason for the question mark next to the "#" symbol meaning the aortic leaflet. PP – right atrium, the remaining signs – as in the previous figures*
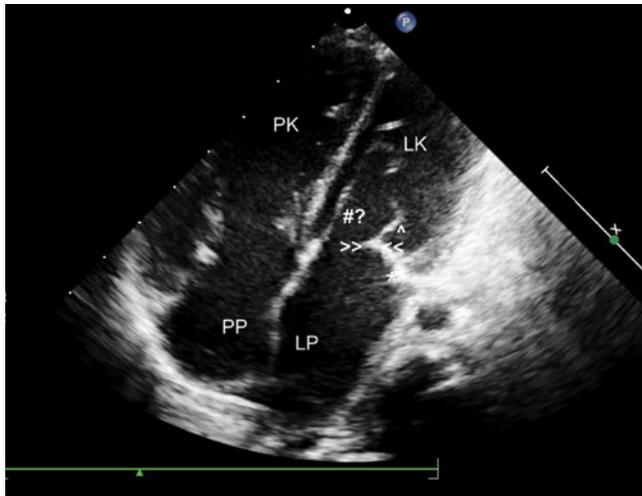
Fig. 7. *The apical four chamber view, slightly modified in reference to Fig. 6: the ultrasound beam has been moved slightly upwards and to the front. This cross-section does not show the connection between the pathological membrane and the aortic mitral leaflet any more. The signs – as in Fig. 6*

Fig. 8. *Parasternal view demonstrating the left ventricle in the transverse axis. In comparison with Fig. 2 the ultrasound beam has been moved slightly towards the atria. Visible linear structure of significant echogenicity running in parallel to the edge of the mural mitral leaflet (marked with the symbol „>> <<"). Also on this level structures which may correspond to the elements of the anterolateral commissure are not visible. The remaining signs as in the previous figures*

2) the hypoplastic form – usually occurring in the left heart hypoplasia syndrome;
3) the supravalvar ring;
4) the parachute valve.

The most widely known, however very rare, situations of the co-occurrence of the parachute valve and the ring include Shone's complex described in 1963, in which the complex stenosis of the inflow tract to the left ventricle (parachute valve and supravalvar ring) occurs together with the stenosis of outflow tract from the left heart ventricle (*left ventricular outflow tract obstruction*, LVOTO) and the stenosis of the aortic arch[4–6]. The supravalvar mitral ring is an anomaly which occurs very rarely[1], sometimes is subtle and causes diagnostic difficulties. The name of the defect suggests the existence of a pathological excrescence located completely in the left atrium, over the mitral ring. However, there are reports of various spatial relationships of the membrane towards the valve: it may be located inside the atrium (completely over the ring), in the intermediate position (over the mural leaflet and on the atrial surface of the aortic leaflet) or it may be completely connected with the leaflets[2,4]. This last location seems to be the most logical one because the suggested background for the formation of the membrane is incorrect division of the mesenchymal tissue of the pericardial cushions[1,3,5]. In such a form the term "supravalvar ring" refers rather to the location of the stenosis above the functional mitral orifice than to the location of the pathological lesion above the anatomic structures of the mitral valve, therefore some authors use the term 'intramitral ring'[7].

The supravalvar ring need to be distinguished from the cor triatriatum sinistrum[1]. Despite of the apparent resemblance (an obstacle for the inflow from the left atrium to the left ventricle located proximally in reference to the mitral orifice) these defects differ significantly. In case of the cor triatriatum the pathologic membrane which is the consequence of the abnormal combination of the primary pulmonary vein with the left atrium, forms – inside the atrium, always above the orifice of its auricle – a diaphragm with a restrictive opening[1]. The supravalvar mitral ring in turn above all limits the diastolic mobility of the leaflets by reducing the inflow area. This dynamic nature of the stenosis may constitute a diagnostic hint during echocardiography. It is not easy to notice a thin membrane slightly protruding out of the atrial surface of the leaflets, however the significant limitation of the diastolic mobility of the leaflets, with the formation of something like a false, additional valve ring may draw the attention of the person carrying out the examination, which will allow for performing a more detailed analysis necessary for making the correct diagnosis[4–6]. It is also necessary to distinguish the valvar ring from the extremely rare supramitral ridge[7], in case of which the intussusception of the free wall of the left atrium in the immediate vicinity of the mitral ring may imitate the image of the supravalvar ring. This fold usually contains a circumflex coronary artery, however it never causes the hindering of the inflow from the atrium to the ventricle, therefore in never constitutes an indication for a cardiosurgical intervention.

The presurgical diagnosis is extremely important in case of the coexistence with another hemodynamically significant defect, e.g. VSD, which – as in the presented case – constitutes the main indication for cardiac surgical correction[4,5]. The approach through the opening of the anterior wall of the right atrium which is applied for the closing of the ventricular septal defect, does not allow for the surgical visualization of the interior of the left atrium nor of the mi-

Fig. 9. *Suprasternal view demonstrating the aortic arch. Visible elongated distal transverse part of the arch as well as clearly accentuated isthmus marked with the symbol "@". Luk dyst – the distal transverse part of the aortic arch, ltsw – left common carotid artery, ltp – left subclavian artery, ao zst – descending aorta*
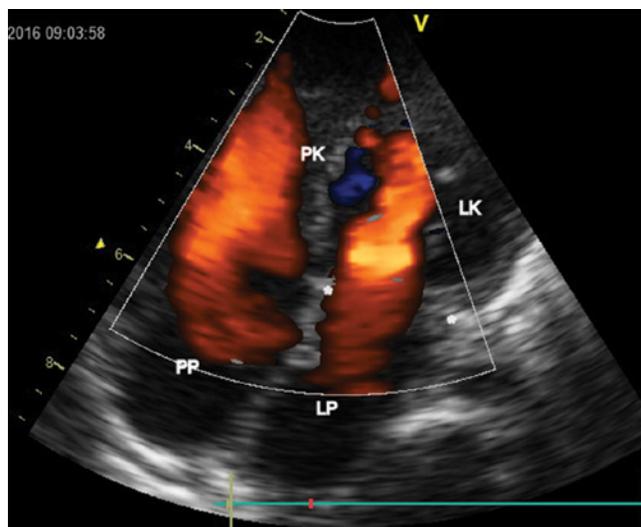
Fig. 10. *Postsurgical examination, the 2nd 24 hours. The apical four chamber view, the inflow from the left atrium to the left ventricle demonstrated using a color Doppler has got a laminar nature. LK – left ventricle, LP – left atrium, PK – right ventricle, PP – right atrium, * – the mitral valve ring*

tral valve and thus does not allow for the intraoperational verification of the diagnosis and for broadening the scope of the intervention. In patients with a supravalvar ring the stenosis has got a progressive nature, therefore over time it contributes to the generation of significant hemodynamic disorders, also in those children in which at the beginning the lesions were not severe[2,5–8]. On the other hand it has to be underlined that even precise resection of the pathological tissue from the atrial surface of the mitral valve leaflets

does not completely normalize their mobility, nor does it fully restore laminar flow which – in the presence of the reduced cross-section of the mitral orifice in case of the parachute valve may lead to the recurrence of the stenosis[5]. The „intravalvar" location of the stenosis which we found in the presented case seems to have worse prognosis in this regard. Therefore patients after supravalvar mitral ring surgery require many years of postoperative supervision[1,3,5,7,8].
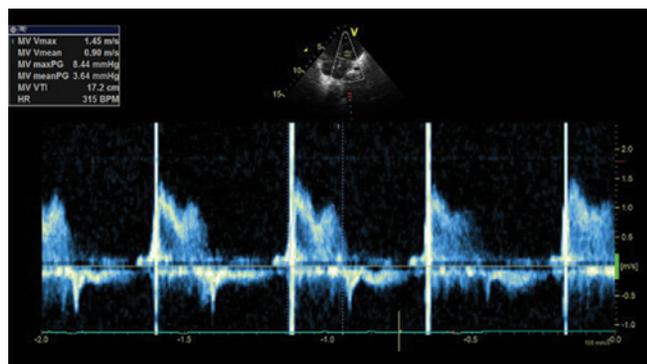
Fig. 11. *Postsurgical examination, the 2nd 24 hours. The measurement of inflow velocity through the mitral valve into the left ventricle. Little residual gradient (maximum 8.44 mm Hg, mean – 3.64 mm Hg) seems to be above all the consequence of the abnormal orifice of the parachute mitral valve – an attempt to cut the leaflets was not made during the correction*

Conclusions

1. In cases of the coexistence of anatomic abnormalities related to the structures of the "left heart" – the mitral valve, the left ventricular outflow tract, the aortic valve, the aortic arch – it is necessary to very carefully analyze the image of the atrioventricular junction because the supramitral membrane may be a very subtle structure which however causes significant hemodynamic disorders.

2. The anatomical image of the described structure is varied and in every case it may have a different, surprising configuration.

Conflict of interest

Authors do not report any financial or personal connections with other persons or organizations, which might negatively affect the contents of this publication and/or claim authorship rights to this publication.

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