**Pediatric Cardiology**

Left Ventricular Outflow Tract Obstruction in Transposition of the Great Arteries. Correlation Between Anatomic and Echocardiographic Findings

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**Introduction and objectives.** Left ventricle outflow tract obstructions in transposition of the great arteries are frequent. We report the correlations between two-dimensional echocardiographic and autopsy findings to draw attention to the usefulness of this diagnostic method in the preoperative evaluation of these anomalies.

**Material and methods.** Of 73 hearts with transposition of great arteries, 26 specimens (38%) with different types of left ventricular outflow tract obstruction were selected to establish the relationship between an anatomical substrate of obstruction and echocardiographic findings in equivalent hearts. Pulsed-wave Doppler studies of velocities at the site of stenosis were done with high-pulse-rate frequency and continuous wave techniques. Eight echocardiographic studies of anatomical specimens and 10 studies in equivalent hearts used for comparison were analyzed to determine correlations.

**Results.** Twenty-eight obstructions were found; the most frequent type being left ventricular outflow tract alteration (77%). The most frequent anomalies were posterior deviation of the infundibular septum, cone-like obstruction, pulmonary valve stenosis and septal hypertrophy, followed by congenital mitral valve anomalies (15%) and anomalies of the tricuspid valve (8%). Two-dimensional echocardiographic studies revealed different types anatomical obstruction.

**Conclusions.** We found precise correlations between the anatomical obstruction and its echocardiographic image. Evaluating electrocardiographic findings is important because these findings can affect the choice of surgical treatment or even the decision to use surgery.

**Key words:** Transposition of the great arteries. Pulmonary stenosis in transposition of the great arteries. Echocardiography. Pathology.

Obstrucciones a la vía de salida del ventrículo izquierdo en la transposición de grandes arterias. Correlación anatomoecocardiográfica

**Introducción y objetivos.** Las alteraciones obstrutivas de la vía de salida del ventrículo izquierdo en la transposición de grandes arterias son frecuentes. Se presenta una correlación anatomoecocardiográfica para resaltar la utilidad de este método diagnóstico en la valoración preoperatoria en este grupo de lesiones.

**Material y métodos.** De un total de 73 corazones con transposición de grandes arterias, se estudiaron 26 especímenes con diferentes tipos de obstrucciones a la vía de salida del ventrículo izquierdo, lo que correspondió al 38%, y que sirvieron de sustrato anatómico para establecer la correspondencia entre la estructura obstructiva y la imagen ecocardiográfica obtenida de corazones equivalentes. El análisis hemodinámico se realizó con ecocardiografía Doppler pulsado, continuo y color. Se revisaron 8 estudios ecocardiográficos que correspondían a las piezas anatómicas y 10 estudios equivalentes para la correlación y análisis.

**Resultados.** Se encontraron 28 obstrucciones; el tipo más frecuente correspondió a las alteraciones propias de la vía de salida del ventrículo izquierdo (77%) y, entre ellas, el desalineamiento posterior del septo infundibular, el cono obstructivo, la estenosis de la válvula pulmonar y la hipertrofia septal, seguidas por las alteraciones de la válvula mitral (15%) y finalmente las alteraciones de la válvula tricúspide (8%). En los estudios ecocardiográficos bidimensionales se reconocieron los diferentes tipos anatómicos de obstrucciones.

**Conclusiones.** La correlación que demuestra este trabajo entre la estructura anatómica obstructiva y su imagen ecocardiográfica fue precisa. La importancia en su valoración radica en que pueden modificar la decisión de tratamiento quirúrgico o incluso hasta contraindicar algún tipo de cirugía.

**Palabras clave:** Transposición de grandes arterias. Estenosis pulmonar en la transposición de grandes arterias. Ecocardiografía. Anatomía patológica.
INTRODUCTION

Surgical treatment for transposition of the great arteries (TGA) requires the assessment of anatomical details that influence the choice of technique used. This is particularly important in patients for whom the best option is anatomical correction. The aim of diagnosis is not only to determine the discordant ventriculoarterial connection, but also to identify any additional alterations caused by the physiopathological peculiarities of left ventricular outflow tract obstructions (LVOTO). Since some of these obstructions are difficult for surgeons to recognize, and since not all are surgically removable (thus surgery is contraindicated), noninvasive, echocardiographic diagnoses confirming their presence and characteristics would be of great help.

The aim of this study was to determine any possible correlation between anatomical and echocardiographic findings to help improve the diagnosis of LVOTOs. The correlation between anatomical obstructions and their ultrasound images was determined with the aim of promoting echocardiography in the preoperative evaluation of these alterations.

MATERIAL AND METHODS

Of a total of 73 hearts with TGA, 26 were found to have different types of LVOTO. The hearts were opened following the normal dissection pattern, although modifications were made in some cases to preserve the obstruction. The samples were classified according to the level of the ventricle where the obstruction was found, and its location in the outflow tract. This previously published classification system1 contemplates four groups (though group IV was not included in this study since it involves the outflow tract of the right ventricle):

–Type I. Alterations of the right ventricular inflow tract that affect the outflow tract of the left ventricle (e.g., prolapse of the septal leaflet of the tricuspid valve due to a perimembranous septal defect), and obstructions caused by overriding of the tricuspid valve.
–Type II. Alterations of the left ventricle inflow tract that affect the outflow tract (e.g., anomalous insertion of the anterior mitral valve chords into the interventricular septum, accessory and ectopic mitral valve tissue, and counter-clockwise positioning of the mitral valve).
–Type III. Alterations of the left ventricular outflow tract and the pulmonary valve, such as deviation of the infundibular septum, hypertrophied funnel-like obstruction and pulmonary valve stenosis.
–Type IV. Alterations of the right ventricular outflow tract (extreme anterior deviation of the infundibular septum).

The anatomical specimens were used to establish the correlation between morphological obstructions and the echocardiographic images obtained with equivalent hearts. Eight echocardiographic studies of the anatomical specimens and 10 studies of equivalent hearts were analyzed to determine any correlations.

The echocardiographic study involved two-dimensional transthoracic and subcostal images complemented by pulsed wave, continuous wave and color power Doppler, following a sequential, segmentary analysis. The abdominal and atrial situs, the pulmonary and systemic venous returns, the type and mode of atrioventricular and ventriculoarterial connections, and the obstructions of the left ventricular outflow tract (as associated lesions) were all defined.

RESULTS

In the 26 anatomical samples with LVOTO, 28 obstructions were found (two obstructions were found in two hearts) (Table 1). The most common types were posterior deviation of the infundibular septum and funnel-like obstruction. The different anatomical types of obstruction were recognizable in the two-dimensional echocardiographic studies.

–Type I. Anomalies of the tricuspid valve. Two anatomical specimens with prolapse of the septal leaflet due to a perimembranous septal defect were examined. Both showed an obstruction in the outflow tract and several degrees of dysplasia of the leaflet (Figure 1a). Echocardiographic studies of equivalent cases (two-dimensional images, parasternal long axis view) showed the valve prolapse that caused the obstruction. The obstruction was quantified with continuous wave Doppler, yielding gradients of up to 30 mm Hg (Figures 1b, c, and d). No cases of obstructive tricuspid valve overriding were seen.
–Type II. Anomalies of the mitral valve. Four specimens (15% of the total material) with mitral valve alterations were studied (one with two associated anomalies). Two showed malpositioning of the mitral valve (one with a ventricular septal defect [VSD]) and associated accessory mitral valve tissue (Figure 2), one with anomalous chord insertions at the border of a ventricular septal defect (Figure 3), and one with

ABBREVIATIONS

TGA: transposition of the great arteries.
LVOTO: left ventricular outflow tract obstruction.
VSD: ventricular septum defect.
accessory mitral valve tissue but with an intact ventricular septum (Figure 4).

Six echocardiograms showing type II obstructions were analyzed: two with mitral malpositioning (one of these was the heart with VDS and associated accessory mitral valve tissue), two showed anomalous insertions of the chords, and the remaining two showed accessory mitral tissue. An echocardiographic view particularly useful to evaluate malpositioning of the mitral valve was the parasternal short axis of the papillary muscles. Observing from the left atrium, clockwise rotation was demonstrated. The altered pattern of the papillary muscles (the anterolateral group of the normal heart) was appreciable. In this anomaly, these structures are found in the anteromedial position, whereas the posteromedial position changes to posterolateral. It is the first of these that causes the obstruction, the chords crossing the outflow tract (Figure 2). This finding was confirmed by an apical five chamber image. The accessory mitral tissue and the anomalous insertion of the chordae tendinae was appreciable in apical two chamber and parasternal long axis views (Figures 3 and 4). The gradients, evaluated in apical views, were not very significant.

–Type III. Alterations in the left ventricular outflow tract and pulmonary valve. These were the most common alterations (77%). The 20 specimens showed 21 obstructions (one heart had two obstructions). Seven showed posterior deviation of the infundibular septum

TABLE 1. Obstructions of the left ventricle outflow tract: 26 anatomical specimens (28 obstructions)

<table>
<thead>
<tr>
<th>Type of obstruction</th>
<th>Number</th>
<th>Total (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type I</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Anomalies of the tricuspid valve</td>
<td>2 (8%)</td>
<td></td>
</tr>
<tr>
<td>Prolapse of the septal leaflet (VSD)</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Overriding</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Type II</td>
<td>4 (15%)</td>
<td></td>
</tr>
<tr>
<td>Anomalies of the mitral valve</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>Accessory tissue</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Malpositioning</td>
<td>2a</td>
<td>2</td>
</tr>
<tr>
<td>Anomalous insertion of chordae tendinae</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Type III</td>
<td>20 (77%)</td>
<td></td>
</tr>
<tr>
<td>Alterations in the left ventricular outflow tract</td>
<td>20</td>
<td>20</td>
</tr>
<tr>
<td>Posterior deviation of the infundibular septum</td>
<td>7b</td>
<td>7b</td>
</tr>
<tr>
<td>Funnel-like obstruction</td>
<td>7</td>
<td>7</td>
</tr>
<tr>
<td>Hypertrophy of the ventricular septum</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>Pulmonary valve stenosis</td>
<td>3</td>
<td>3</td>
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*a Associated with mitral valve accessory tissue. *b Associated with pulmonary valve stenosis.
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Fig. 2. A: Interior view of the left chambers showing the malpositioning of the mitral valve. Note the positioning of the chordae tendinae over the left ventricular outflow tract (asterisk) and the mitral valve accessory tissue (MVAT). B: Echocardiographic images of the same case (parasternal long axis view) showing the insertion of the chord into the anterior papillary muscle (arrow) and the ventricular septal defect (asterisk). C: The diagram shows the abnormal positioning of the mitral valve, whose anterior commissure is located below the pulmonary valve. The position of the posteriomedial and anteromedial papillary muscles show the clockwise rotation as seen from the atria, or the counter-clockwise rotation as seen from apex. D: echocardiogram showing cross section of the papillary muscles. A clockwise rotation of the mitral valve is seen from the left atrium, the papillary muscles ending up in a posteriomedial and anteromedial position. The latter causes the obstruction.

MVAT indicates mitral valve accessory tissue; LML, lateromedial leaflet; MML, mitral medial leaflet; PM, posteriomedial; AL, anteriolateral; PL, posteriolateral; AM, anterioesfial; LV, left ventricle; RV, right ventricle; PA, pulmonary artery.

Fig. 3. A: Internal view of the left ventricle (LV) showing the anomalous insertion of a mitral valve (MV) on the ventricular septum at the lower edge of the interventricular communication (VSD). B: Diagram showing an obstruction caused by insertion of the chords into the septum. C: Echocardiographic images showing the anomalous insertion of a mitral chord (arrow) into the interventricular septum, giving rise to this slight obstruction. Note also the VSD and the connection of the left ventricle with the pulmonary artery. D: site of obstruction observed with color power Doppler.

LV indicates left ventricle; MV, mitral valve; VSD, ventricular septal defect; PA, pulmonary artery; LA, left atrium; Nao, neoaoorta.
(all with VSD, one associated with pulmonary valve stenosis), seven funnel-like obstructions (all with VSD), three cases of hypertrophy of the interventricular septum (intact septum), and three with pulmonary valve stenosis (all with VSD) (Figures 5 and 6).

The five echocardiographic studies included: one patient with posterior deviation of the infundibular septum (demonstrated from the parasternal long axis), one with funnel-like obstructions (Figure 5), one with hypertrophy of the septum and integral septum, and two with pulmonary valve stenosis (Figure 6). In this last type of obstruction, the short parasternal axis of the great vessels allowed the pulmonary valve with its thickened leaflets to be examined. Fixed echocardiographic images of the patient with posterior deviation of the infundibular septum were not demonstrative and are not shown.

**DISCUSSION**

LVOTOs are seen in hearts with normal connections between the cardiac segments as well as in those with complete TGA. In the latter, they are caused by several mechanisms. The literature reports the frequency of these lesions to vary between 30% and 35%; in the present material this frequency was slightly higher.
(38%, i.e., 28 obstructions in 26 specimens since autopsy material was used. Obstructions are divided into either dynamic or anatomical (fixed) types. The dynamic forms are more commonly associated with an intact ventricular septum. Anatomical types should be suspected when there is a VSD, in which case it should be differentiated from the dynamic forms and their exact nature determined.

Several types of anatomical obstruction have been described, both with and without VSD. The classification used in the present study^4^ includes all the fixed forms mentioned above; this systematic arrangement makes it possible to understand where the obstruction is in the outflow tract. Type I includes anomalies of the tricuspid valve, such as prolapse of the septal leaflet due to VSD (favored by the greater pressure of the right ventricle and generally accompanied by dysplasia of the valve,^6^ as seen in the anatomical specimens). The overriding of the tricuspid valve is obstructive when the inlet VSD extends anteriorly and includes a perimembranous component, and when the chords insert into the papillary muscle of the left ventricle, crossing part of the outflow tract.

Type II corresponds to anomalies of the mitral valve,^7^-^9^ the majority associated with VDS and without functional significance, including accessory mitral valve tissue, the anomalous insertion of one or more chords, and malpositioning of the valve. This last alteration should be distinguished from a mitral cleft, which can be confused with a commissure in the malpositioning of the mitral valve. In a true cleft, unlike in a commissure, there are no chords or papillary muscles in the cleft region near the valve ring. This can be discerned echocardiographically. With respect to mitral malpositioning, unlike in the above communications, the valve is rotated in a clockwise fashion when observed from the left atrium, or counterclockwise when observed from the apex of the left ventricle. Echocardiography adequately defined the position of the papillary muscles. This group should include subvalvular stenoses described as membranes since one of their formation mechanisms is related to alterations in the development of the anterior leaflet of the mitral valve (in the majority of cases), giving rise to fibrous ridges running from the mitral ring to the membranous septum. In patients with an intact interventricular septum, subvalvular obstructions caused by a fibrous ring have been reported. This ring is analogous to that seen in slight subaortic stenosis in patients with ventriculoarterial concordance.
Echocardiographically it is difficult to tell whether the subaortic fibrous ring is formed by alterations of the mitral valve or by true fibromuscular protrusions of the interventricular septum into the left ventricular outflow tract. Information supporting the existence of a true subaortic fibrous ring would be the lesion’s more regular shape plus an origin in the septum close to the valve plane; mitral alterations are more irregular. However, proper differentiation lies at the histological level. Although mitral alterations made up 15% of the specimens studied, the literature suggests that this anomaly is more common than normally expected, appearing in up to 22% of patients with TGA.

Type III corresponds to alterations of the outflow tract and of the pulmonary valve. In this investigation, specimens with posterior deviation of the infundibular septum and funnel-like obstruction were more common than previously reported. In this group, alterations of the pulmonary valve can be echocardiographically distinguished. The valve shows dysplasia with thickened, fused sigmoid leaflets, causing steeper gradients. In septal hypertrophy it is difficult to distinguish whether the obstruction is dynamic or fixed. Echocardiographic information that helps differentiate this group from the dynamic obstructions includes the bulging of the septum towards the left ventricle, mesosystolic closure of the pulmonary valve, and systolic anterior movement of the mitral valve. In addition, the left ventricle has thin walls. The fixed forms cause the left ventricle to show hypertrophy to different degrees.

As mentioned above, the majority of anatomical or fixed LVOTOs are accompanied by VSD. In some, such as the posterior deviation of the infundibular septum, funnel-like obstruction or prolapse of the tricuspid septal leaflet, the defect forms part of the alteration. In the present material, the obstructive forms with intact interventricular septa included fibromuscular septal protrusion and isolated pulmonary stenosis. Malpositioning of the mitral valve, accessory mitral material, and anomalous insertion of the mitral chords may –or may not– be associated with VSD.

These obstructions are usually in isolation, but on occasion, a single heart can show more than one type, as observed in the anatomical material examined. This means care must be taken when making a diagnosis. It is notable that in patients with TGA there are many mechanisms that lead to LVOTO, as this study shows. This is in contrast to that seen in obstruction of the right ventricle in hearts with normal connections between the cardiac segments. It is possible, therefore, that in TGA there is some still undetermined mechanism that favors the development of these lesions.

The importance of properly diagnosing this type of TGA-associated anomaly lies in the fact that some are surgically unresectable or might be complicated by certain techniques, e.g., the Rastelli procedure should not be used in prolapse of the tricuspid septal leaflet (although it is thought that placing the interventricular patch as a wedge could separate the septal leaflet from the tricuspid and embrace the aorta). Anatomical correction would be contraindicated in some cases, e.g., when there is anomalous insertion of the mitral chords or when this valve is malpositioned: the mitral valve would later have to support systemic pressure. From a surgical point of view, the most relevant mitral anomalies are the displacement of the ring and/or of the tensor apparatus associated with the VSD, in which, association with a cleft mitral valve is common. A dysplastic or stenotic pulmonary valve would require a review of the surgical techniques used. Although pulmonary stenosis can be repaired, it is often insufficient, which is not desirable after anatomical correction. Frequently, the valvular gradient can be overestimated given the increase in pulmonary blood flow, especially when there is a wide arterial conduit. Generally, tricuspid anomalies, accessory mitral tissue and a subpulmonary membrane can be resected and condition no residual obstruction. However, posterior deviation of the infundibular septum can cause a residual obstruction and even contraindicate anatomical correction.

The correlation between the anatomical and echocardiographic findings found in this study show the degree of sensitivity of this technique. Echocardiography is no longer just for confirming a diagnosis or assessing left ventricle function. However, LVOTOs are not very evident and usually generate gradients of little significance, so they must be sought out to give surgeons more complete and more precise information. Particularly helpful in the detection of these alterations are the parasternal long axis view, the apical view with anterior angulation to assess the left ventricular outflow tract, and subcostal views. The anomalies of the atroventricular valves are better visualized in apical two and four chamber views, the long and short parasternal view, and the subcostal four chamber view. Pulsed and continuous wave Doppler studies help locate the obstruction and aid in the quantitative estimation of the gradient. Color power Doppler shows the extension of a stenosis. An obstruction is said to exist if the Doppler-measured gradient is greater than 20 mm Hg.

CONCLUSIONS

This study shows a precise correlation between anatomical obstructions and their echocardiographic images, underlining the sensitivity of this diagnostic method. These associated anomalies, which are often difficult to visualize, should be intentionally sought out during assessment of patients with TGA in order to provide more complete and precise information to surgeons. This is important since some obstructions
cannot be surgically resected; for some types, surgery may even be contraindicated.

The obstructive mechanisms of the left ventricle outflow tract in patients with TGA are several, in contrast to that seen in hearts with normal intersegmental connections. There may therefore be a mechanism that favors the development of these lesions in patients with the former problem.

REFERENCES