The optimal procedure for the great arteries and left ventricular outflow tract obstruction. 
An anatomical study☆

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Abstract

Objective: To describe the optimal surgical strategy in heart specimens with transposition of the great arteries (TGA) and left ventricular outflow tract obstruction (LVOTO). Methods: Thirty-three specimens with LVOTO were selected: TGA with intact ventricular septum (TGA/IVS) (10), TGA/VSD (21), and Taussig—Bing (2). Results: LVOTO in TGA/IVS consisted of combinations of bicuspid pulmonary valve (four), subpulmonary fibrous ridge (four), obstructive muscular conus (two) and bulging muscular septum (four). Arterial switch operation (ASO) with LVOTO resection/valvotomy was feasible in nine hearts. Obstructive anterior papillary muscle prohibited LVOTO relief in one specimen. In TGA/VSD and Taussig—Bing LVOTO consisted of combinations of bicuspid (nine) or unicommissural (one) pulmonary valve, fibrous ridge (three), obstructive muscular conus (five), malaligned outlet septum (six), accessory mitral valve tissue (two), straddling mitral valve (two) and anterior mitral valve rotation (four). VSDs were subpulmonary in 13 (9 perimembranous, 4 muscular), subaortic in 3 (2 perimembranous, 1 anterior muscular), doubly committed in 2, inlet in 3 (2 perimembranous, 1 muscular), non-committed and anterior in 1, and finally 1 VSD extended both into inlet and subpulmonary outlet septum. LVOTO resection and ASO with VSD closure was possible in 10. In six specimens, both a Rastelli and a Nikaidoh operation were feasible. For two hearts, a Nikaidoh procedure was the only option, while Rastelli was considered optimal in another specimen. Mitral valve anomalies prevented LVOTO relief in four, only permitting for Senning/VSD closure (one) or univentricular palliation (three). Conclusions: LVOTO resection and pulmonary valvotomy frequently permits an ASO. Inlet VSD, impossibility of VSD enlargement, straddling mitral valve, distant aorta and small right ventricle make the Nikaidoh procedure the best option. Mitral anomalies preventing LVOTO relief can make biventricular repair impossible.

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Keywords: Transposition great arteries; Pulmonary stenosis; Arterial switch operation; Nikaidoh operation; Rastelli operation; REV operation

1. Introduction

The anatomy of pulmonary stenosis in discordant ventriculo-arterial connection may vary considerably. The pulmonary valve can be bicuspid (seldom unicommisural) and obstructive. Subvalvular obstruction may be caused by a discrete fibrous ridge or membrane, by accessory tissue from an atroventricular valve, or from the perimembranous region. Malattachment or straddling of the mitral valve is sometimes present and can result in left ventricular outflow tract obstruction (LVOTO). Muscular forms of LVOTO include an anterolateral muscle (ALM) bundle, leftward deviation of the infundibular septum, and the presence of subpulmonary muscular conus. Tunnel-form LVOTO is usually associated with hypoplasia of the pulmonary valve annulus.

A bicuspid and obstructive pulmonary valve is sometimes amenable to valve repair after which an arterial switch operation (ASO) may offer a good solution [1,2]. Valvular stenosis by fused commissures in a bicuspid pulmonary valve can be treated by valvotomy in the same way as in a stenotic bicuspid aortic valve and should not be considered an absolute contraindication for an ASO.

Also, subpulmonary obstructive tissue can many times be resected allowing for an ASO [1,3]. Subpulmonary stenosis in the form of a fibrous ridge or membrane can be enucleated, while posteriorly deviated septal muscle can usually be resected. In case of a hypoplastic pulmonary annulus or non-resectable LVOTO, an ASO is impossible. Repair can be performed by different procedures: the Rastelli procedure, the Réparation à l'Etage Ventriculaire (REV) operation, the Metras modification, or the operations that were described by Bex and Nikaidoh. An atrial rerouting procedure (Senning/Mustard) may be an option depending on the degree of
pulmonary stenosis that will remain. A LV-to-pulmonary artery (PA) conduit has been described but is not a desirable option [1,3—12]. The anatomy usually allows for biventricular repair, but in some hearts only univentricular palliation is possible. Straddling of the tricuspid valve, or, especially, of the mitral valve, may prohibit ventriculopulmonary septation.

We describe a series of cardiac specimens with transposition of the great arteries (TGA) and LVOTO obstruction, either with or without VSD. The method of sequential segmental analysis and description of all associated anomalies was used for this purpose. After careful and detailed description of the specimens, we aim to evaluate the different surgical options in each specimen. Pros and cons of the different procedures are analyzed and reported in relation to the specific anatomy of each specimen.

The anatomical details of many of the specimens in this study have been described previously [13—16]. However, new surgical techniques have been developed since, and we aim to analyze the anatomical details of the specimens in relation with the current surgical techniques.

2. Materials and methods

The selected specimens were repeatedly and independently studied and described by a fellow in pediatric cardiac surgery, a pediatric cardiac surgeon, and a cardiac anatomist. After having described all anatomical details, the hearts were studied again in order to determine the best surgical options for each specimen.

2.1. Anatomical criteria

The Leiden collection comprises more than 2500 specimens with congenital cardiac anomalies, most of which have not been operated. From the 200 specimens with TGA we have selected 33 specimens with TGA and important LVOTO. Hearts with other-than-concordant atrioventricular connections, specimens with importantly unbalanced ventricles, and specimens with double outlet right ventricle (RV) were excluded from this study. DORV hearts were not included as DORV was considered as forming a too heterogeneous group of diagnoses for the purpose of this study. An exception was made for two specimens (3646, 4538) with TGA and subpulmonary VSD that showed overriding of the pulmonary artery (Taussig—Bing malformation with LVOTO). Specimens in which surgery had changed the anatomy severely were also excluded. For these reasons, not all specimens that are mentioned in earlier publications were selected for this study; on the contrary, some more recent heart specimens have been added [13—16].

2.2. Surgical criteria

Several considerations were used for this purpose. Generally speaking, if an ASO was possible this was considered to be the best choice. Resection of LVOTO and/or repair of a stenotic pulmonary valve were considered if these procedures would allow for an ASO (±VSD closure) without remaining serious LVOTO and without risk of early pulmonary (neo-aortic) valve replacement [1—3]. The Nikaidoh operation was preferred over REV, Metras and Rastelli procedures. REV and Metras procedures were preferred over the Rastelli operation [7]. The procedure of aortic translocation that was originally described by Bex et al. can be considered as being a good alternative to the Nikaidoh procedure; the Bex procedure can be used in patients with TGA and LVOTO with intact septum. In the description by Bex the outlet septum was not resected and the first reported patient had no VSD [5]. Relief of LVOTO can be adequately performed through the pulmonary valve, but full resection of the outlet septum may, in some instances, be the only way to completely desobstruct the LVOT. Leaving the RV in the systemic circulation (after an atrial switch procedure) was judged to be less optimal. An atrial switch procedure with a LV-to-PA conduit was thought to be an undesirable choice. Orthotopic placement of a RV-to-PA connection or conduit (as in the Nikaidoh procedure) was preferred over an extracardiac positioned RV-to-PA conduit (as in the Rastelli procedure), which always includes a right ventriculotomy [4,6,7,8]. A direct LV-to-aorta connection was considered to be better than an oblique or angulated tunnel between LV and aorta. Special attention was given to a possible decrease in volume and function of the RV after surgical repair. Procedures that would lead to a diminished RV volume were considered as being not optimal. Positional anomalies of the atrioventricular valves were in some instances considered to be an important obstacle to septation and biventricular repair. Finally, it was assumed that the coronary anatomy would never form an obstacle for either an ASO or a posterior translocation of the aorta.

3. Results

3.1. Anatomy

Of the 33 selected specimens, 10 had TGA with intact ventricular septum (TGA/IVS), 21 had TGA with VSD, and 2 had Taussig—Bing anomaly.

3.1.1. TGA/IVS/LVOTO

Four pulmonary valves were bicuspid, and LVOTO was caused by a fibrous ridge or tunnel (four), obstructive muscular conus (two), bulging of septal muscle (four), or anterior malattachment or malrotation of the mitral valve (one). Discontinuity of the mitral and the pulmonary valve resulted in subpulmonary muscular conus and was responsible for different degrees of LVOTO obstruction. The presence of an ALM bundle was responsible for obstructive bulging of muscular tissue in six specimens. Combinations of obstructive anatomic substrates were frequent (Table 1).

3.1.2. TGA/VSD/LVOTO

Nine pulmonary valves were bicuspid and one was unicommisural. Subpulmonary obstruction was caused by a fibrous ridge or tunnel in three and by an obstructive muscular conal tunnel in five. Malalignment of the outlet septum toward the LV was observed in six specimens. Obstruction by anomalies of the mitral valve was encountered in eight specimens: accessory mitral valve tissue in
Table 1
Anatomical characteristics and suggestions for surgical repair in specimens with TGA/IVS and L VOTO

<table>
<thead>
<tr>
<th>Number</th>
<th>Basic anomaly</th>
<th>L VOTO</th>
<th>Best procedure</th>
<th>Second best procedure</th>
<th>Position great arteries</th>
<th>Coronary anatomy</th>
</tr>
</thead>
<tbody>
<tr>
<td>1959</td>
<td>TGA/IVS</td>
<td>Bicuspid pulmonary valve</td>
<td>ASO with valve repair</td>
<td>—</td>
<td>Aorta right anterior</td>
<td>1L-2CxR</td>
</tr>
<tr>
<td>5686</td>
<td>TGA/IVS</td>
<td>Bicuspid pulmonary valve</td>
<td>ASO with valve repair</td>
<td>—</td>
<td>Aorta right anterior</td>
<td>1Lcx-2R</td>
</tr>
<tr>
<td>2804</td>
<td>TGA/IVS</td>
<td>Dynamic muscular obstruction</td>
<td>ASO</td>
<td>—</td>
<td>Aorta right anterior</td>
<td>1Lcx-2R</td>
</tr>
<tr>
<td>2780</td>
<td>TGA/IVS</td>
<td>Fibrous ridge, bulging outlet septum</td>
<td>ASO with resection</td>
<td>—</td>
<td>Aorta right posterior</td>
<td>1R-2LCx</td>
</tr>
<tr>
<td>4225</td>
<td>TGA/IVS</td>
<td>Fibrous ridge</td>
<td>ASO with resection</td>
<td>—</td>
<td>Aorta right anterior</td>
<td>1Lcx-2R</td>
</tr>
<tr>
<td>5540</td>
<td>TGA/IVS</td>
<td>Bicuspid pulmonary valve and fibrous ridge</td>
<td>ASO with resection</td>
<td>—</td>
<td>Aorta right anterior</td>
<td>1L-2CxR</td>
</tr>
<tr>
<td>5557</td>
<td>TGA/IVS</td>
<td>Bicuspid pulmonary valve, fibrous ridge and bulging septum</td>
<td>ASO with resection</td>
<td>—</td>
<td>Aorta right anterior</td>
<td>1L-2CxR</td>
</tr>
<tr>
<td>5986</td>
<td>TGA/IVS</td>
<td>Obstructive muscular conus and ALM</td>
<td>ASO with resection</td>
<td>—</td>
<td>Side by side</td>
<td>1R-2LCx</td>
</tr>
<tr>
<td>6682</td>
<td>TGA/IVS</td>
<td>Anterior mitral malrotation and malattachment with anterior papillary muscle. Accessory mitral tissue</td>
<td>Senning/Mustard</td>
<td>Univentricular</td>
<td>Side by side</td>
<td>1RL-2Cx</td>
</tr>
</tbody>
</table>

LVOTO: left ventricular outflow tract obstruction; TGA/IVS: transposition of the great arteries with intact ventricular septum; ASO: arterial switch operation; ALM: anterolateral muscle.

two, straddling of the mitral valve in two, and (anterior) rotation of the mitral valve with septal attachment of the anterior papillary muscle in another four. Obstructive lesions of the LVOTO were also frequently found in combination with each other (Table 2).

3.1.3. VSD

In 13 specimens the VSD was mostly committed to the pulmonary valve (9 perimembranous subpulmonary VSDs, 4 muscular subpulmonary VSDs). In two hearts the VSD was equally committed to the aortic and the pulmonary valve (in the presence of an outlet septum) and was therefore defined as being doubly committed. In three specimens the VSD was more related to the aortic valve (perimembranous subaortic in two and muscular/subaortic subaortic in another one). These VSDs were located more anteriorly than the VSDs related to the pulmonary valve. A fourth anteriorly located muscular VSD did not have an evident commitment to either the aortic valve or the pulmonary valve. In three hearts the VSD was mainly located in the inlet septum (perimembranous inlet in two and muscular inlet in one). Finally, one large perimembranous inlet VSD also extended into the outlet septum and showed commitment to the pulmonary valve (Table 2).

3.1.4. Position great arteries/coronary anatomy (TGA/IVS and TGA/VSD)

The position of the great arteries was as follows: aorta right anterior to the pulmonary artery in 18, aorta side by side to the pulmonary artery in 8, aorta directly anterior in 4, aorta right posterior in 2, and aorta left anterior to the pulmonary artery in 1 specimen (with associated dextrocardia). The coronary anatomy was classified according to the Leiden classification [15]. 1LCx-2R was found in 17 specimens, 1L-2CxR was observed in 5, 1RL-2Cx in 6, and 1R-2LCx in 4 hearts. One specimen presented with a single coronary ostium (2LCxR). A double LAD system was present in three hearts. All but one heart with 1LCx-2R or 1L-2CxR coronary anatomy had the aorta anterior or right anterior to the pulmonary artery. In one specimen with 1LCx-2R anatomy and dextrocardia the aorta was positioned left anteriorly to the pulmonary artery. In all hearts with 1RL-2Cx or 1R-2LCx anatomy the aorta was either positioned side by side or right posteriorly to the pulmonary artery (Tables 1 and 2).

3.2. Surgery

3.2.1. TGA/IVS/LVOTO

In all but one heart with TGA/IVS an ASO was considered to be possible. All four bicuspid pulmonary valves were judged to perform well after an ASO: no or minimal insufficiency and no or minimal obstruction. In two bicuspid valves a limited commissurotomy was considered necessary to optimize valve function.

Discrete fibrous ridges were detected in four specimens and could all easily be resected. An anterolateral muscle (muscle of Moulaert) was found in two specimens and occurred in association with obstructive subpulmonary muscular conus [17]. In both hearts resection of the obstruction through the pulmonary valve was considered to be possible, supposedly resulting in an unobstructed LVOTO.

In one specimen LVOTO was caused by an anteriorly rotated mitral valve that was not repairable. An ASO (or a Bex procedure) would have resulted in unacceptable LVOTO. However, an atrial switch procedure (Senning or Mustard) with resection of the obstructive accessory fibrous tissue would result in some RVOT obstruction that was judged to be acceptable. The alternative would have been univentricular palliation.

3.2.2. TGA/VSD/LVOTO

In 10 specimens an ASO with VSD closure and LVOTO resection was judged feasible. Accessory fibrous tissue attached to the mitral valve presented in two specimens and could be resected resulting in an unobstructed LVOTO (Fig. 1). In one specimen LVOTO was caused by an aneurysmatic membranous septum protruding into the LVOT. This aneurysmatic tissue could be resected or tightened. In six hearts resection of obstructive muscular tissue was deemed necessary for relief of LVOTO. Obstruction by malalignment of the outlet septum was present in three of these specimens. Resection of (part of) the outlet septum would lead to an unobstructed LVOTO. Resection could always be performed through the pulmonary valve.

In two hearts repair of a bicuspid pulmonary valve was needed to perform an ASO. In one specimen mild hypoplasia of the RV and the tricuspid valve was observed. However, this was judged to be compatible with an acceptable outcome following ASO.
<table>
<thead>
<tr>
<th>Number</th>
<th>Basic anomaly</th>
<th>VSD</th>
<th>LVOTO</th>
<th>Best procedure</th>
<th>Alternative procedure</th>
<th>VSD enlargement necessary with Rastelli?</th>
<th>Comment</th>
<th>Position great arteries</th>
<th>Coronary anatomy</th>
</tr>
</thead>
<tbody>
<tr>
<td>181</td>
<td>TGA/VSD/LVOTO</td>
<td>Perimembranous subpulmonary</td>
<td>Obstructive, dysplastic, bicuspid pulmonary valve; annular hypoplasia</td>
<td>Nikaidoh</td>
<td>REV/Rastelli</td>
<td>Yes</td>
<td>REV/Rastelli will result in a small RV</td>
<td>Aorta right anterior</td>
<td>1LCx-2R</td>
</tr>
<tr>
<td>435</td>
<td>TGA/VSD/LVOTO</td>
<td>Perimembranous subpulmonary</td>
<td>Obstructive, dysplastic, bicuspid pulmonary valve; annular hypoplasia; straddling mitral valve; cleft anterior mitral leaflet</td>
<td>Univentricular pathway</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>884</td>
<td>TGA/VSD/LVOTO</td>
<td>Perimembranous subpulmonary</td>
<td>Straddling mitral valve; cleft anterior mitral leaflet</td>
<td>Univentricular pathway</td>
<td></td>
<td></td>
<td></td>
<td>Aorta anterior</td>
<td>1LCx-2R</td>
</tr>
<tr>
<td>3337</td>
<td>TGA/VSD/LVOTO</td>
<td>Perimembranous subpulmonary</td>
<td>ALM; posteromedian muscle; conus</td>
<td>ASO with VSD closure and resection of obstructive muscle tissue</td>
<td></td>
<td></td>
<td></td>
<td>Side by side</td>
<td>1RL-2LCx</td>
</tr>
<tr>
<td>3570</td>
<td>TGA/VSD/LVOTO</td>
<td>Perimembranous subpulmonary</td>
<td>Anteroseptal twist; hypertrophy of the LV anterior wall</td>
<td>ASO with VSD closure and resection of obstructive muscle tissue</td>
<td></td>
<td></td>
<td></td>
<td>Aorta right anterior</td>
<td>1LCx-2R</td>
</tr>
<tr>
<td>6134</td>
<td>TGA/VSD/LVOTO</td>
<td>Perimembranous subpulmonary</td>
<td>Anterior mitral valve rotation and malalignment of the anterior papillary muscle; incomplete cleft anterior mitral leaflet</td>
<td>Senning with VSD closure and residual LVOTO</td>
<td>Univentricular pathway</td>
<td>n.a.</td>
<td>Both Nikaidoh and REV/Rastelli impossible as the LVOTO cannot be relieved without damage to the mitral valve</td>
<td>Aorta right anterior</td>
<td>1LCx-2R</td>
</tr>
<tr>
<td>6198</td>
<td>TGA/VSD/LVOTO</td>
<td>Perimembranous subpulmonary</td>
<td>Bicuspid obstructive pulmonary valve; anterior mitral malattachment</td>
<td>Nikaidoh</td>
<td>REV/Rastelli</td>
<td>?</td>
<td>REV/Rastelli will result in a small RV</td>
<td>Aorta right anterior</td>
<td>1L-2CxR</td>
</tr>
<tr>
<td>5554</td>
<td>TGA/VSD/LVOTO</td>
<td>Perimembranous subpulmonary</td>
<td>Bicuspid pulmonary valve; fibrous ridge; anterior mitral malattachment</td>
<td>Nikaidoh</td>
<td></td>
<td>n.a.</td>
<td>REV/Rastelli impossible due to mitral malattachment on outlet septum</td>
<td>Aorta right anterior</td>
<td>1LCx-2R</td>
</tr>
<tr>
<td>3646</td>
<td>TGA/DOR/LVOTO</td>
<td>Perimembranous subpulmonary</td>
<td>Accessory mitral valve tissue</td>
<td>ASO with VSD closure and resection of accessory tissue</td>
<td></td>
<td>n.a.</td>
<td></td>
<td>Side by side</td>
<td>1R-2LCx</td>
</tr>
<tr>
<td>3268</td>
<td>TGA/VSD/LVOTO</td>
<td>Muscular subpulmonary</td>
<td>Aneurysmatic tissue of membranous septum</td>
<td>ASO with VSD closure and resection of aneurysmatic tissue</td>
<td></td>
<td>n.a.</td>
<td></td>
<td>Aorta right anterior</td>
<td>2LCxR (single coronary ostium)</td>
</tr>
<tr>
<td>3371</td>
<td>TGA/VSD/LVOTO</td>
<td>Muscular subpulmonary</td>
<td>Bicuspid dysplastic pulmonary valve; malalignment outlet septum</td>
<td>ASO with VSD closure and resection of outlet septum; shaving of the pulmonary valve</td>
<td></td>
<td>n.a.</td>
<td></td>
<td>Aorta right anterior</td>
<td>1LCx-2R</td>
</tr>
<tr>
<td>3856</td>
<td>TGA/VSD/LVOTO</td>
<td>Muscular subpulmonary</td>
<td>Pulmonary annulus hypoplasia; obstructive conus</td>
<td>Nikaidoh</td>
<td>REV/Rastelli</td>
<td>Yes</td>
<td>REV/Rastelli will result in a small RV</td>
<td>Aorta right anterior</td>
<td>1L-2LCxR</td>
</tr>
<tr>
<td>4538</td>
<td>TGA/DOR/LVOTO</td>
<td>Muscular subpulmonary</td>
<td>Fibrous ridge; ALM</td>
<td>ASO with VSD closure and resection of fibromuscular tissue</td>
<td></td>
<td>n.a.</td>
<td></td>
<td>Side by side</td>
<td>1RL-2LCx</td>
</tr>
<tr>
<td>4731</td>
<td>TGA/VSD/LVOTO</td>
<td>Doubly committed</td>
<td>Unicommissural pulmonary valve; malalignment outlet septum</td>
<td>Nikaidoh</td>
<td>REV/Rastelli</td>
<td>Yes</td>
<td>REV/Rastelli will result in a small RV</td>
<td>Aorta left anterior (dextrocardia)</td>
<td>1LxR-2Cx (quadricuspid aortic valve)</td>
</tr>
<tr>
<td>4402</td>
<td>TGA/VSD/LVOTO</td>
<td>Doubly committed</td>
<td>Bicuspid pulmonary valve; annular hypoplasia</td>
<td>Nikaidoh</td>
<td>REV/Rastelli</td>
<td>No</td>
<td>Both Nikaidoh and REV/Rastelli possible</td>
<td>Aorta right anterior</td>
<td>1LCx-2R</td>
</tr>
<tr>
<td>6679</td>
<td>TGA/VSD/LVOTO</td>
<td>Perimembranous subaortic anterior</td>
<td>Obstructive conal tunnel; bicuspid pulmonary valve</td>
<td>REV/Rastelli</td>
<td></td>
<td>No</td>
<td>REV/Rastelli will give straight connection LV-Ao</td>
<td>Side by side</td>
<td>1RL-2Cx</td>
</tr>
<tr>
<td>5760</td>
<td>TGA/VSD/LVOTO</td>
<td>Perimembranous subaortic</td>
<td>Malalignment outlet septum</td>
<td>ASO with VSD closure and partial resection of outlet septum</td>
<td></td>
<td>n.a.</td>
<td></td>
<td>Side by side</td>
<td>1RL-2Cx</td>
</tr>
</tbody>
</table>
In the two hearts with Taussig–Bing anomaly the subaortic RVOT was also considered as being (potentially) narrow. To prevent later RVOT obstruction part of the outlet septum and muscle bundles from outlet septum to the anterior RV free wall would need to be resected.

In another nine hearts with TGA/VSD and L VOTO biventricular repair was possible, but pulmonary valve hypoplasia and/or non-resectable subpulmonary obstruction restricted the use of an ASO.

In these specimens the surgical options were either Rastelli/REV procedure or Nikaidoh operation. In two specimens Rastelli or REV procedures were not possible while a Nikaidoh operation was judged to be technically feasible with good outcome. Anterior rotation of the mitral valve inhibited creation of a LV-to-aorta tunnel but after resection of the obstructive pulmonary valve and the complete outlet septum the aorta could be detached and moved posteriorly on top of the LVOT. The anomalous position of the mitral valve would not result in L VOTO. In the second specimen extension of the VSD into the inlet septum was considered to impair tunneling the LV to the aorta. The tunnel would obstruct the RV inlet (Fig. 2A and B). Therefore, a Nikaidoh procedure was judged to be the only technique to accomplish biventricular repair.

In five specimens the Rastelli procedure was considered to result in a small RV and, therefore, a Nikaidoh operation was thought to be a better way of repair (Fig. 3). One of these specimens (4731) had a unicommissural pulmonary and a quadricuspid aortic valve, and has been described earlier [18].

In two specimens (4402, 6679) the Rastelli operation was not thought to result in important reduction of RV volume and similar considerations could be made for REV procedure. In specimen 6679 a Nikaidoh procedure was considered as not leading to a better position (more on top of the LV) of the aorta in relation to the LV. The VSD here was perimembranous, subaortic, anterior, and large. The aortic valve was big and the LVOT very small. Moving the aorta over a very limited distance would not have meant any improvement in the position of the tunnel LV-aorta. Only the presence of an extracardiac RV-to-PA conduit would be a negative point for the Rastelli operation in these two specimens.
In four specimens LVOTO could not be adequately relieved. In one specimen (6134) with anterior rotation of the mitral valve and anterior papillary muscle malattachment the LVOTO was considered to be sufficiently mild to perform an atrial switch operation with a RVOT gradient afterwards. Repositioning of the mitral valve was not possible, and the option of leaving the malpositioned mitral valve in the LVOT following an ASO was considered as undesirable. Univentricular management of the anomaly was considered to be the only option in three hearts with straddling atrioventricular valves. In one specimen (2915) with a tricuspid valve straddling through a big inlet VSD, septation was not possible, thus blocking the way to biventricular repair. In two other specimens (435, 884) the mitral valve straddled through a subpulmonary VSD. Septation was judged to be impossible and univentricular palliation would be the only choice (Fig. 4).

4. Discussion

4.1.1. TGA/IVS/LVOTO

In one specimen with TGA/IVS malrotation of the mitral valve prohibited an ASO. In all other specimens an ASO was considered to result in an acceptable outcome when combined with repair of a bicuspid pulmonary valve or resection of subpulmonary stenosis. As is true for a bicuspid aortic valve in normal hearts, a bicuspid pulmonary valve after an ASO in TGA may need later repair or replacement, especially when valvular dysplasia was present and/or a commissurotomy was necessary initially. However, the mere presence of a bicuspid pulmonary valve is not a contraindication for an ASO. The incidence of reoperation is generally reported to be low [1–3]. Of the 311 patients who have received an ASO in Leiden University Medical Center, 10 had an isolated bicuspid pulmonary valve (3.2%). No patient has now more than grade I insufficiency and no significant transvalvular gradients are observed at last follow-up. No patient was reoperated for dysfunction of the bicuspid pulmonary valve. However, follow-up of the arterial switch operation in our institution is maximally 30 years and bicuspid pulmonary valves in TGA may degenerate at a later stage.

A discrete fibrous ridge in the LVOT is sometimes, although infrequently, observed in TGA/IVS. At the time of the ASO the fibrous ridge can easily be resected or enucleated. The presence of a fibrous ridge indicates an abnormal anatomy or
flow profile of the LVOT and, similar to fibrous subaortic stenosis in normal hearts, there will be a chance of recurrence of this fibrous LVOTO even after successful enucleation [1,3]. A dynamic muscular LVOT obstruction can be present in TGA/IVS and will usually resolve after the ASO when the RV/LV pressure ratio is normalized [19]. In TGA there is normally a fibrous connection between the pulmonary and the mitral valve. In case of muscular conus the pulmonary and the mitral valves are separated by muscular tissue. An anterolateral muscle may be present and is typically located between the anterior leaflet of the mitral valve and the anteroseptal wall of the LV [17]. In these cases, the mitral valve may be positioned a little more posteriorly and slightly rotated [13]. In our series of TGA/IVS this was observed in specimens 5986 and 5540. An ALM can usually be resected through the pulmonary valve allowing for an unobstructed LVOT.

Thus, for TGA/IVS the risks of late dysfunction of the pulmonary valve and reoperation for LVOTO recurrence appear to be sufficiently low to consider an ASO with LVOTO resection/pulmonary valve repair as the best possible solution [1–3].

4.1.2. TGA/VSD/LVOTO

Deviation of the outlet septum always results in a malalignment VSD and to obstruction of usually the RVOT. In TGA, deviation of the outlet septum through the VSD may, however, lead to considerable obstruction of the LVOT situated either anterior to the pulmonary orifice or posterior as part of subpulmonary conus (Fig. 5). Therefore, LVOTO is more frequent and usually more severe in TGA with VSD than it is in TGA/IVS.

In a high number (9 of 23) of specimens an ASO with VSD closure and LVOTO resection was found to be technically feasible. Accessory tissue originating from the mitral valve and aneurysmatic tissue of the membranous septum can be resected with no or minimal chance of recurrence (Fig. 1). Care should be taken not to damage the atrioventricular valves, as the accessory valve tissue is sometimes difficult to distinguish from the valve tissue itself. Muscular obstruction in the form of muscular subpulmonary conus, an ALM, or a malaligned outlet septum can be resected, opening the way for an ASO. In our opinion, whenever an ASO can be performed, this should be considered the treatment of choice.

When the pulmonary valve is hypoplastic or when the LVOTO cannot be resected completely, the LV needs to be connected to the aorta to allow for biventricular repair. The VSD may have to be enlarged to create an unobstructed communication. VSD augmentation can be done by resecting the outlet septum, as has been described by Lecompte et al [6]. The VSD can also be enlarged by resection of part of the trabecular septum in the postero-inferior corner of the VSD. To facilitate a LV-to-aorta connection the VSD must, at least, have some relation to the aorta. Inlet septal defects or more apically located muscular VSDs are generally not suitable for the Rastelli or REV procedures. In our specimens we found that six times both a Nikaidoh operation and a Rastelli/REV operation could be performed. However, two times a Nikaidoh procedure was the only way to construct a biventricular heart: a Rastelli procedure could not be performed because of malposition of the mitral valve or because of extension of the VSD into the inlet septum (Fig. 2A and B). In the six specimens where both Nikaidoh and Rastelli procedures were possible, we observed five times that significant RV volume reduction after the Rastelli procedure would occur (Fig. 3). It can be postulated that a REV operation in some of these hearts would result in slightly less RV volume reduction as the VSD patch can be made somewhat straighter than in the Rastelli procedure [7].

When comparing the procedures, the theoretical advantages of the Bex—Nikaidoh procedure or posterior translocation of the aorta are obvious: no volume reduction of the RV, an ‘anatomical’ connection of LV to aorta, orthotopic placement of the RV-to-PA conduit, and no right ventriculotomy. Aortic translocation is the only technique that permits biventricular repair in case of inlet or distant muscular VSD, straddling mitral or tricuspid valve, or an already small RV. The detachment of the aortic root from the RV and resection of the outlet septum permit an optimal exposure for ventricular septation, which can be especially helpful in the presence of malpositioned or straddling atrioventricular valves. In the procedure that was described first by Bex et al., the posterior aortic translocation was not combined with resection of the outlet septum and resection by a patch as was described by Nikaidoh et al. [5,9]. The original Bex procedure is probably a good solution for patients with TGA and LVOTO with a small VSD or with intact septum as long as the LVOTO can be desobstructed fully through the pulmonary valve.

The Bex and the Nikaidoh procedures are technically more demanding and involve the risks associated with coronary transfer and aortic valve detachment and reinsertion. We have assumed for the purpose of this study that coronary artery transfer would never pose a problem, neither for an arterial switch procedure nor for a Bex or a Nikaidoh operation. However, some coronary artery anatomies may increase the risk of coronary artery transfer. Detachment of the aortic root from the right ventricle may be more hazardous with a right coronary artery springing from sinus 1 and running close to anterior part of the aortic root.

![Fig. 5. Specimen 3403: opened LV with malalignment VSD (anterior) with deviation of the outlet septum through the VSD toward the LVOT. The subpulmonary Infundibulum is imprisoned by the outlet septum together with the ALM, the mitral valve, and the ventricular septum. PO: pulmonary orifice; ALM: anterolateral muscle.](https://academic.oup.com/ejcts/article-abstract/31/5/879/358974)
Increasing experience with the arterial switch operation, the Ross and Konno operations provide the technical skills that are necessary for posterior aortic translocation [9–12]. Several authors have reported that the long-term results of the Rastelli procedure are not satisfactory. The incidence of late mortality is high as is the number of reoperations. Replacement of the extracardiac conduit is needed in almost all cases, and there appears to be an important incidence of reoperation for residual or recurrent LVOTO [20, 21]. RV-to-PA homografts in extracardiac position have been reported to be less durable than homografts in an orthotopic pulmonary position [22]. For the abovementioned reasons, the Lecompte/Metras procedures and the Nikaidoh operations should probably be given priority over the classical Rastelli operation in many patients with TGA, VSD, and LVOTO.

Finally, in some hearts, univentricular palliation can be the only surgical option. Ventricular septation may not be possible because of straddling of the mitral or the tricuspid valve. Straddling valves do not always inhibit VSD closure: sometimes the straddling chords can be detached from the septum and reinserted after suturing the VSD patch. However, in the specimens we have examined this would either be impossible or very difficult as straddling chords could not be resected and reattached without severe damage to valvular function. Furthermore, we found a cleft anterior leaflet to be frequently associated with straddling mitral valve, which would have complicated valve reconstruction even more (Fig. 4). In another study of anatomical specimens, this not-so-infrequent association of TGA and mitral valve anomalies has been described earlier by Moene and Oppenheimer-Dekker [16]. Especially in the presence of LVOTO the anatomy of the mitral valve should therefore be exactly clarified before proceeding with surgical repair.

The limitation of this study is that the specimens form a selection that may not be fully representative for clinical practice. Furthermore, some degree of surgical subjectivity was unavoidable when deciding upon the best procedure for each specimen.

References


Appendix A. Conference discussion

Dr. D. Metras (Marseille, France): We all know that a switch operation with a transposition and normal bicuspid valve is currently done. Now, you say that examining your specimens you could think that with the repair of unicommissural or bicuspid pulmonary valve, which probably is stenotic, you could do switch. Does that mean that presently if you find this situation you would do a switch operation?

Dr Hazeemp: Well, I did say that in the group with VSD we found a unicommissural pulmonary valve that was in the subgroup where the annulus of the pulmonary valve was very small and we had to do another option, a Nikaidoh, or Rastelli operation. So this was not amenable to arterial switch operation. And I think that a unicommissural pulmonary valve is not a good idea to do a switch operation.

Dr Metras: But even with a bicuspid with stenosis, you would do a switch operation?

Dr Hazeemp: Depending on the degree of valve abnormality. If a small valvotomy would be sufficient to relieve the problem, then probably we would go on with a switch operation.
**Dr W. Mrowczynski (Poznan, Poland):** I wanted to ask from which era do your specimens come from?

And what is the relation between the data you show and your real clinical life, I mean the spectrum of TGAs you encounter?

**Dr Hazekamp:** The specimens come from long time ago when the rate of autopsies was a lot higher than it is nowadays. So it’s not representative for what we are doing in the clinic. So you should not make a relation between the clinical situation right away.