Biventricular repair of double outlet right ventricle with non-committed ventricular septal defect (VSD) by VSD rerouting to the pulmonary artery and arterial switch

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Abstract

Objectives: Biventricular repair of double outlet right ventricle non-committed ventricular septal defect (DORVncVSD) is usually achieved by a VSD rerouting to the aorta. This technique can be limited by the presence of tricuspid chordae and by the pulmonary artery to tricuspid valve distance. Furthermore, there is an important risk of late subaortic obstruction related to the long patch required that creates a potential akinetic septal area. Presented here is another technique; by VSD rerouting to the pulmonary infundibulum and arterial switch.

Methods: Ten patients, with DORVncVSD, underwent a VSD rerouting to the pulmonary infundibulum followed by arterial switch. Seven had a previous pulmonary artery banding and one a moderate infundibular stenosis. The median age at surgery was 16 months (range 3 weeks to 4.5 years). All patients had a bilateral infundibulum, with a large persistent subaortic conus, D malposition of the aorta, side-by-side vessels and double loop coronary patterns. The VSD was perimembranous with inlet or trabecular extension. The operation was conducted through a combined approach. The VSD was constantly enlarged superiorly. The almost permanent subaortic obstruction was released. The VSD was always found quite close to the pulmonary infundibular ostium. The arterial switch technique was adapted to the complex coronary anatomy.

Results: There was one non-cardiac death. At a mean follow-up of 20 months, all nine survivors are in NYHA class I, in sinus rhythm, and have no subaortic gradient greater than 15 mm. Conclusion: This technique of VSD rerouting to the pulmonary artery and arterial switch limits greatly the size of the rerouting patch, respects the tricuspid chordae and is independent of the pulmonary artery–tricuspid valve distance. In this early series of biventricular repair of DORVncVSD, the VSDs were always found close to the pulmonary artery, allowing this new type of repair. © 2002 Elsevier Science B.V. All rights reserved.

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1. Introduction

Double outlet right ventricle (DORV) is mainly characterized by a malposition of the aorta that arises from the right ventricle and represents a primitive mode of ventriculo-arterial connection [1]. Depending on the location of the ventricular septal defect (VSD) and the degree of malposition of the aorta, the patients with DORV present with cardiopathies that have similar hemodynamics than VSD, Fallot’s tetralogy or transposition of the great arteries (TGA). DORV with non-committed VSD (DORVncVSD) represents the extreme end of the anatomical spectrum. Its biventricular repair remains a high risk procedure [2]. The important distance between the VSD and the remote aorta implies to construct a long prosthetic intra cardiac tunnel that can impair not only the tricuspid and the pulmonary valves but also the left ventricular (LV) function in creating a significant akinetic area. This tunnel presents a high risk of obstruction during growth and is a serious limitation of the technique. Despite the uncertainties of the long-term results of the Fontan operation [3], many groups have been so far faithful to the univentricular palliation, that represents a less risky alternative [4].

The aim of this article is to present another surgical option with VSD rerouting to the pulmonary infundibulum...
and arterial switch, that seems almost always feasible in DORVncVSD in the absence of valvar pulmonary stenosis.

2. DORV anatomy

2.1. DORV definition

The recent Surgical Nomenclature adopted by the Data Bases of the STS and EACTS [10,11] and also by the AEPC [12] defines four types of DORV:

- DORV with subaortic VSD
- DORV Fallot-type, with subaortic or doubly committed VSD and pulmonary outflow stenosis
- DORV TGA-type (Taussig Bing), with subpulmonary VSD
- DORV non-committed VSD

The definition of DORV remains controversial. If there is unanimity around the 50% rule given by Anderson and colleagues [5,6], other definitions [7–9] remain valuable. The presence of a subaortic conus, as stated by Van Praagh et al. [8], remains the major landmark of this anomaly. Lecompte has proposed to change the term DORV for malposition of the great arteries [9].

2.2. DORVncVSD definition

The term non-committed VSD was introduced by Lev and Barati [7]. Van Praagh et al. [8] used the term non-committed VSD and mentioned the ‘considerable’ distance between the VSD and the aorta. These terminologies have been quite vague and a precise definition was needed. Barbero-Marcial et al. [13] define it as a VSD either muscular inlet without perimembranous connection or a VSD of atrioventricular (AV) canal type. Belli et al. [14] define it as a VSD distant from the arterial valves by a distance superior to an aortic diameter; this definition includes perimembranous VSD.

The definition (Figs. 1–3) that we propose includes:

1. A VSD distant from both aortic and pulmonary annulus by a length superior to an aortic diameter
2. Both great vessels arising 200% from the right ventricle
3. Constantly, a double conus

This definition excludes:

The Taussig Bing heart where the VSD, located above the trabecula septomarginalis [16], is close to the pulmonary annulus by a distance inferior to an aortic diameter and where the pulmonary artery does not arise 100% from the right ventricle

The DORV with AVSD where the VSD is extended
below the aorta, which represents probably the majority of this condition

This definition includes:

The DORVncVSD with RVOT stenosis either infundibular or valvular and the forms with pulmonary atresia
The rare forms of DORV-AVSD where the VSD is distant from the aortic annulus
The DORVncVSD with single functioning ventricle, although they will not be described here as not being compatible with biventricular repair.

2.3. Anatomical features of DORVncVSD (Figs. 1–4)

The presence of a double infundibulum is definitely the major landmark [8,9]. The ‘aortic infundibulum’ being separated from the pulmonary infundibulum by a conal septum that does not contribute to the ventricular septation. Both great vessels are totally (200%) on the right ventricle and this is a clear landmark of the DORVncVSD. The VSD is typically a perimembranous VSD (Figs. 1 and 2) with inlet or trabecular extension; sometimes it can be strictly perimembranous. The VSD is located beneath the posterior limb of the trabecula septomarginalis (Fig. 2), in contact with the tricuspid annulus, as demonstrated by Freedom and Yoo [16]. The VSD can be restrictive with a diameter less than an aortic diameter. Others [13] have described strictly muscular VSD: either inlet, mid-trabecular or apical, with intact perimembranous septum. A long-lasting pulmonary artery (PA) banding can induce a VSD restriction. The vessels are usually side by side, the aorta being frequently posterior. The aorta has a very abnormal location, being maintained very high on the right ventricle; it stands partly out of the myocardium with the sinus of Valsalva well visible. In fact, the important distance between the VSD and the aorta is not in relation with the location of the VSD but is essentially due to the very abnormal and remote location of the aorta that sits at the top of the RV.

The distance between the tricuspid and pulmonary annuli is a crucial landmark for biventricular repair, as outlined by Lecompte [9], because the aortic tunnel is to be constructed in this area. The major problem is the frequent presence of conal tricuspid chordae that stand in the way of the ventricular tunnel to be constructed [9,13,14,16].

The coronary distribution (Fig. 4) shows an abnormal course with double looping [17], with a posterior location of the main trunk and a right coronary crossing anteriorly the aorta. Frequently, there is a single coronary right ostium giving the left main trunk and the RCA.

2.4. Associated lesions

Among the associated lesions, the subaortic obstruction is more a permanent lesion than an association. Almost constantly, one or several abnormal subaortic muscular bands is found that connect the right part of the aortic infundibular wall to the conal septum (Fig. 5B). A real DORVncVSD with AVSD seems quite rare; usually the AVSD is a type C of Rastelli with a large subaortic component of the VSD [18] that is therefore committed to the aorta. Nevertheless, AVSD with a VSD not extended
below the aorta has been described in two patients with DORV by Paci fi co et al. [19] and in one patient by Mee and colleagues [20].

Pulmonary stenosis can be associated; it is often a muscular obstruction with normal pulmonary valve and annulus. Elsewhere, it is a valvar stenosis. Straddling AV valves can be present and also overriding, the latter being a contraindication to biventricular repair. Criss-cross heart can be also associated.

Moderate LV hypoplasia allows biventricular repair. True single functioning ventricles are frequent and excluded from the study.

3. Patients

From January 1998 to April 2001, ten patients, with DORVncVSD, underwent a biventricular repair that included a VSD rerouting to the pulmonary infundibulum followed by an arterial switch. The median age at surgery was 16 months, ranging from 3 weeks to 4.5 years. Three patients were less than 1 year old, including one neonate. There were six boys and four girls. Seven patients had a previous PA banding and one a moderate infundibular stenosis. The diagnosis was obtained by echocardiography and eight patients had an angiography (Fig. 5).

All patients had a bilateral infundibulum, with a large persistent subaortic conus, D malposition of the aorta, side-by-side vessels and double loop coronary patterns. There were four patients with single coronary coming from the right ostium (Fig. 4). The VSD was perimembranous with inlet or trabecular extension. It was restrictive with an interventricular gradient greater than 20 mm in four patients. Subaortic obstruction was constant. The VSD was located far away from both the arterial valves by a distance greater than the aortic diameter (Fig. 5).

The associated lesions included: four ostium secundum, two non-connected LSVC, two restrictive mitral valve with normal subvalvular apparatus, two moderate LV hypoplasia with LV end-diastolic volume at 16 and 18 ml/m^2, one muscular infundibular stenosis, two tricuspid valve straddling of type A and one left juxtaposition of appendages.

4. Methods

All operations but one were performed by the same surgeon. Five patients were operated at Marie Lannelongue Hospital in Paris, one at the Eppendorf University Hospital in Hamburg, Germany, one at Fu Way Hospital in Beijing, China, one at the Bakulev Institute in Moscow, Russia; one at the Queen Silvia Hospital in Gothenburg, Sweden, and one at the Institute Cuore Morgani in Catania, Sicily, Italy.

The operation was performed on full-flow cardiopulmonary bypass at moderate hypothermia with repeated crystalloid cardioplegia. The mean cross clamping time was 122 ± 21 min, ranging from 95 to 185 min. It was conducted through a triple approach; through the tricuspid valve, the pulmonary infundibulum and the aortic annulus. The RV incision was performed on the low part of the pulmonary infundibulum. A double exploration through the tricuspid valve and the infundibulum allows location of the VSD; it was always found in contact with the tricuspid valve and very close to the pulmonary infundibulum. The distance between the tricuspid valve and the conal septum was reduced in six patients. In five patients, tricuspid...
control was obtained in seven patients showing no signifi-
cant LV outflow tract (LVOT) gradient. One patient
pre- sented a moderate mitral valve stenosis, two patients
have moderate aortic regurgitation and one patient presents
a RVOT stenosis with 40 mmHg gradient.

6. Discussion

6.1. Anatomy

DORV anatomy remains probably the most controversial
entity on congenital heart diseases. The reason is probably
that DORV is not a disease but only a primitive mode of
ventriculo-arterial connection.

The embryology is becoming a more precise science with
the enormous interest brought by the use of embryonic cells.
The introduction of immunohistochemistry in cardiac
embryology research has allowed precise statements [1].
Using monoclonal antibodies targeted against different anti-
gens expressed by the embryological tissues (GIN2), it is
possible to follow the migration of cardiomyocytes along
the embryological development [15]. These recent publica-
tions confirm that the primitive right ventricle is a DORV
containing the conotruncus. The migration of the aorta
toward the mitral valve is a complex procedure, named
wedging, that allows the aortic side of the conotruncus to
nestle between the tricuspid and the mitral valve [1]. These
elements explain partly why the right ventricular structures
are usually normal in DORV, this primitive RV bearing
abnormally an additional structure: the entire conotruncus,
including the aortic infundibulum, the infundibulum septum
and the pulmonary infundibulum.

In DORVncVSD, the major landmark is the entire location
of the great vessels on the right ventricle. To follow the
Anderson rule, one can say that the great vessels are 200% on
the RV. The entire conotruncus is on the RV including an
aortic and a pulmonary conus separated by a conal septum
that has nothing to do with the ventricular septation. The
other forms of DORV contain only partially the great vessels;
the Taussig Bing contains only 50% of the PA and the
DORV-Fallot 50% of the aorta. These forms should better
be named TGA or Fallot, respectively. It would be probably
tsimpler to reserve the name DORV for patients with 200%
vessels on the right ventricle, implying a double conus.

The definition of DORVncVSD is also controversial. The
‘considerable’ distance [8] between the VSD and the two
arterial valves needs to be quantified. Belli et al. [14] have
proposed that in DORVncVSD, this distance should be
greater than the aortic annulus diameter. If this definition is
appropriate, it seems more precise to add two other condi-
tions: 200% vessels on the right ventricle and a double conus.

The VSD, in DORVncVSD, is located below the trabecula
septomarginalis (Fig. 3) as pointed out by Freedom and Yoo
[16]. This allows to differentiate the DORVncVSD from the
Taussig Bing hearts were the VSD is located above the TSM
and therefore, close to the pulmonary annulus.

The definition given by Barbero-Marcial et al. [13] relies
only on the location of the VSD that should be muscular,
located in the inlet or trabecular septum without perimem-
branous extension; excluding all connections with the tricus-
pid valve. In fact, the VSD is usually perimembranous as
mentioned by Becker and Anderson [5] and Freedom and
Yoo [16], and as it was found in all of the patients in this
series.

The ‘considerable’ distance between the VSD and the
great vessels is not due to an abnormal location of the
VSD but is related to a very abnormal location of the
aorta that is standing at the roof of the right ventricle. There-
fore, it is more a remote aorta than a remote VSD that is a
cause of DORVncVSD.

It is noticeable that in DORVncVSD, the right ventricular
structures are in place; namely, the tricuspid valve, the
infundibular septum and the pulmonary artery (Figs. 1–3).
The disorder is created by the additional presence of the
entire aortic conus that has remained in place as in primitive
hearts. The stability of the right ventricular structures in
DORVncVSD explains why the VSD is usually located
next to the ostium infundibuli, making possible the new
technique described.

Finally, the real anatomical anomaly of the DORV is a
malposition of the aorta, the right structures being in place. Lecompte and Sidi [9] have proposed the term ‘malposition of the great vessels’ that is partly correct. The term ‘malposition of the aorta’ seems more appropriate and describes the real anomaly that is an abnormal remainder of the aortic conus.

6.2. Surgery

The intraventricular tunnel rerouting of the VSD to the aorta is the operation of reference for DORVncVSD. In the series of 21 patients reported by Belli et al. [14], the overall mortality is 10% without late death. In two patients, the repair was abandoned during the operation because of conal tricuspid chordae. There have been six reoperations for late subaortic stenosis (29%) and no late deaths.

In the series of 18 patients reported by Barbero-Marcial et al. [13], the early mortality is 11% with no late subaortic stenosis but there were three late deaths, including two sudden deaths. An elegant multiple patch technique to prevent subaortic obstruction is presented. According to the authors, the presence of abnormal tricuspid chordae was not a contraindication and could be resolved by either reimplantation of the tricuspid chordae on the patch or sacrifice of one leaflet and bicuspidization of the tricuspid valve. A reduced pulmonary-tricuspid distance [23] was also not a contraindication, and the pulmonary valve was sacrificed and a valved conduit inserted.

The real risk of this technique, in addition to the potential impairment of the tricuspid and pulmonary valves, is the mandatory presence of a very long patch into the RV. The risk of subaortic obstruction is evaluated at 30% by Belli et al. [14,21,22] and seems to be correlated with the length of the patch. Furthermore, this long patch creates an important akinetic area in the LVOT obstruction that can impair the LV function.

The other option is the Fontan operation, preferred by many centers [3,4]. In comparison with the potential risks of the VSD to aorta rerouting, a Fontan, undertaken here on two ventricles, remains a satisfactory option. In cases of type AV valve straddling, overriding or severe LV hypoplasia, it is the only option.

The rerouting to the PA followed by arterial switch is our preferred solution. The short length of the tunnel is a key issue and the longer the tunnel, the greater the risk of late subaortic obstruction. Our early experience allows confirmation that when the VSD is very far away from the aorta, it is almost always close to the ostium infundibuli (Figs. 1 and 2). The first advantage of this technique is to construct a shorter tunnel (Figs. 3 and 6), reducing the risk of subaortic obstruction. The second advantage is that the rerouting to the PA is independent of the tricuspid valve to PA distance and that it is not involved with the tricuspid chordae (Fig. 6). Interestingly, in our series the VSD was always perimembranous. Barbero-Marcial et al. [13] exclude these forms in the definition of DORVncVSD. Further studies are required to clarify this point; our opinion is that an intact perimembranous septum is extremely rare in DORV for embryological reasons and that the VSD is not committed in DORVncVSD, mainly because of the remote position of the aorta. The arterial switch requires translation of coronary arteries with double loop coronary courses, side-by-side vessels and frequent single coronary. These types of transfer are now well controlled [17]. They require, nevertheless, an adapted technique, the pulmonary bifurcation needing to be transferred towards the right PA [17] to prevent the compression of the left-sided coronary trunk, in cases of associated Lecompte maneuver. The arterial switch is therefore complex and may require a second learning curve in comparison to a normal coronary course switch.

7. Conclusion

Biventricular repair of DORV with non-committed VSD remains a challenging procedure that is based on a precise evaluation of the anatomical features. The major distance between the aorta and the VSD is more in relation to a very abnormally high location of the aorta that sits at the roof of the right ventricle. The major landmark of the anomaly is the persistence of an important subaortic conus that represents an addition to otherwise normal right ventricular structures. The DORVncVSD is essentially a malposition of the aorta.
The intraventricular tunnel connecting the VSD to the aorta is the repair of reference. This technique seems nevertheless quite aggressive and is associated with an important risk of late subaortic obstruction when the VSD is very distant.

Our early experience, in a series of ten patients, with rerouting of the VSD to the ostium infundibuli followed by arterial switch, is satisfactory with one non-cardiac death and no late subaortic obstruction. It is our operation of choice when the aorta is very distant. This technique creates a smaller tunnel, is not involved by the presence of conal tricuspid chordae, and is independent of the distance between the pulmonary and the tricuspid valve.

References


Appendix A. Conference discussion

Dr J. Fragata (Lisbon, Portugal): The vessels were side by side in all patients. Why did you perform the LeCompte maneuver? Do you actually need to do that? Or is that to release tension from the coronary anastomosis?

Dr Lacour-Gayet: Well, when you don’t perform the LeCompte maneuver, there is a better relationship of the vessels. But you can never resolve the coronary relocation. When relocating the right coronary button posteriorly, you take a risk that this button is squeezed between the two vessels, particularly in this condition with frequent single coronary artery, where the left trunk runs behind the pulmonary artery. My experience has been to remain quite faithful to the LeCompte maneuver. Now, I know that others have reached a satisfactory result without the LeCompte maneuver.

Dr C. Tchervenkov (Montreal, QC, Canada): I don’t intend to get into the discussion we had yesterday about complete AV canal and DORV. I would strictly like to focus on the fact that this technique is extremely innovative and offers an excellent solution for that subgroup of patients with non-committed VSD and DORV. However, Francois, you very well know that there are patients that have significant either valvular or subvalvar pulmonary stenosis, and some patients have VSD that is completely confined to the inlet such as in complete AV canal and DORV. Have you encountered any such patients in your series, and how would you deal with these patients?

Dr Lacour-Gayet: When the patient has an RVOT stenosis, if the stenosis is located at the site of the pulmonary valve, this is a contraindication for this technique. If it is located at the site of the infundibulum, this can be resected or patched and it is not a contraindication. If the operation of biventricular repair implies a sacrifice of the tricuspid and the pulmonary valve and construction of a very large tunnel inside the right ventricle, I think that the option of univentricular repair is more satisfactory.

As to your last question, regarding the AV canal and DORV, our experience is not in favor of a frequent association between DORV and the AV canal; these patients in our experience have been for the great majority of Fallot’s tetralogy with AV canal, with a subaortic component of the VSD.