Abnorma ventriculoarterial connections in situs solitus and atrioventricular concordance: relations between outlet septum trabecula septomarginalis and ventriculo infudibular fold

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ABSTRACT

Objective: To evaluate the relations between Outlet Septum, Trabecula Septomarginalis and Ventriculoinfundibular Fold as landmarks for classifications of abnormal Ventriculoarterial Connections and related surgical approaches.

Methods: Observations on 33 hearts.

Results: The relations between Outlet Septum, Anterior and Posterior Limb of the Trabecula Septomarginalis and Ventriculoinfundibular Fold characterize the different forms of Ventriculoarterial Connections. In Tetralogy of Fallot and in Double Outlet Right Ventricle with subaortic Ventriculooseptal Defect the Antero Superior Limb inserts into the Outlet Septum displaced anteriorly. In Double Outlet Right Ventricle with subpulmonary Ventriculooseptal Defect the Postero Inferior Limb inserts into the Outlet Septum which may fuses with the Ventriculoinfundibular Fold. In Double Outlet Right Ventricle with absence of Outlet Septum the Postero Inferior Limb inserts into the Ventriculoinfundibular Fold. When the pulmonary valve originates mostly from the left ventricle the Ventriculoarterial Connections are Discordant and the Posteroinferior Limb inserts into the Ventriculoinfundibular Fold.

Conclusions: The Ventriculoarterial Connections may be described with sequential-segmental approach by relations between Outlet Septum, limbs of Trabecula Septomarginalis and Ventriculoinfundibular Fold.

Key Words: Double-Outlet Right Ventricle, Outlet Septum, Trabecula Septomarginalis, Ventriculoinfundibular Fold.

RESUME

Objectif: étudier le relations entre Septum Infundibulaire, Trabécule Septomarginale et Replie Ventriculo Infundibulaire a visée de la classification et des indications chirurgicales dans la pathologie de la Connexion Ventriculaire Artérielle

Méthodes: observations sur 33 cœurs.

Résultats: Les relations entre Septum Infundibulaire, Trabécule Septomarginale et Replie Ventriculo Infundibulaire entraînent les différentes morphologies pathologiques de la Connexion Ventriculaire Artérielle. Dans la Tétralogie de Fallot et dans le Ventricle Droit à Double Issue avec Communication Interventriculaire sous aortique c’est la Branche Antérieure qui rejoint le Septum Infundibulaire tandis que dans le Ventricle Droit à Double Issue et Communication Interventriculaire sous pulmonaire c’est la Branche Postérieure qui rejoint le Septum Infundibulaire et peut se fondre avec le Replie Ventriculo Infundibulaire. Dans le Ventricle Droit à Double Issue avec absence du Septum Infundibulaire la Branche Postéro Inférieure rejoint le Replie Ventriculo Infundibulaire. Lorsque plus de la moitié de la valve pulmonaire est supportée par le ventricule gauche on parle de Connexion Ventriculaire Artérielle Discordante et la Branche Postéro Inférieure rejoint le Replie Ventriculo Infundibulaire.

Conclusions: chaque pathologie de la Connexion Ventriculaire Artérielle peut être décrite de façon sequentielle et segmentaire par les relations entre Septum Infundibulaire, Trabécule Septo Marginale et Repli Ventriculo Infundibulaire.

Mots-clés: Ventricle Droit à Double Issue, Septum Infundibulaire, Trabecule Septomarginale, Repli Ventriculo Infundibulaire.

1. Introduction

There is a spectrum of cardiac malformations known as “Cono Truncal Anomalies” which reflect an abnormal transfer during development of the aorta from the right to the left ventricle. R H Anderson proposed a system of nomenclature and classification of Congenital Heart Disease totally descriptive with sequential-segmental analysis according to the sequential approach of R. Van Praagh [1-2]. There is a wide variation of Infundibular Morphology and Arterial Relationships which occur with any type of Ventriculoarterial Connection: these conditions should neither be named nor defined in terms of arterial relationships and infundibular morphology. The segmental connections are the hallmark of the
lesions. The International Nomenclature [3-4] adopted by the databases of the Society of Thoracic Surgeons (STS), the European Association of Cardiothoracic Surgery (EACTS) and also by the Association for European Pediatric Cardiology (AEPC), define four types of Double-Outlet Right Ventricle (DORV) based on the clinical presentation and treatment:

1: Ventricular Septal Defect (VSD).
2: Tetralogy of Fallot (TF).
3: Transposition of Great Arteries (TGA).
4: DORV with non committed Ventricular Septal Defect (VSD).

A unified nomenclature facilitate the creation of a Cardiac Surgery Database. However, a prerequisite for successful cardiac surgery is understanding cardiac anatomy using words in their accepted sense and avoiding artificial conventions [5]. The sequential-segmental approach provides a description of complex anomalies such as the Cono Truncal Anomalies with all its variations. It is not a matter of semantic pedantry but introducing precision to and removing ambiguity. This will improve the understanding of practitioners and subsequently benefit the patients.

The key feature in the morphology is the location and insertion of the Outlet (or Infundibular) Septum (OS) to the Ventricular Septum (VS) and the position of the Ventriculoseptal Defect (VSD).

In this sequence, the structure of the Inner Curvature of the heart separating the attachments of atrioventricular and arterial valves (Ventriculo Infundibular Fold, VIF) is also important, as is the extent of the Septomarginal Trabeculation (Trabecula Septo Marginalis, TSM) with the Anterosuperior Limb (AL) and the Posteroinferior Limb (PL). We will show that the Ventriculoarterial Connections in all types of Cono Truncal Malformations with atrioventricular concordance may be characterized by the relationships between OS, TSM, VIF and VSD so as to understand the optimal applications of different surgical procedures. In view of the rarity of other varieties of Double Outlet Right Ventricles (DORV), this review is limited to DORV with Situs Solitus, Atrio Ventricular Concordance and normally or near normally sized ventricles (suitable for biventricular repair).

Fig 1 (a, b): These dissections of a normal heart show the relationships of TSM (septal band) and the supraventricular crest. (a) Dissection in which the roof of the right ventricle has been removed. It shows that the greatest part of the supraventricular crest is the inner curvature of the parietal wall of the right ventricle (VIF). Further dissections (b) reveals that part of this fold forms the freestanding subpulmonary infundibulum (Sub-pulm. infundibulum). There is no “outlet septum” as such identified in the normal heart. Note also the location of the triangle of Koch, seen well in (a).

Fig 2: In this specimen the muscular outlet septum is deviated posteriorly relative to the crest of the ventricular septum producing subpulmonary stenosis. The ventriculoarterial connections are discordant (complete transposition).
these relationships defining the anatomy of the IS, OS, VIF, TSM and the VSD throughout the spectrum of conotruncal malformations anticipating to which morphology each surgical principle can be better applied.

**Biventricular correction**

In this spectrum of malformations there are two main options for definitive biventricular repair: intraventricular re-routing and the arterial switch operation. Some hearts can be corrected simply by closing the VSD so that the aorta is re-routed to the left ventricle [6]. In others, an intracavitary tunnel must be created within the right ventricle to achieve this effect [7-8-9]. For side-by-side relationship of the great arteries, the intraventricular re-routing is the preferable method but it is not always possible to predict whether an unobstructed subaortic route can be created from the left ventricle to the aorta. Resection of the OS between the subaortic route and VSD is mandatory with reattachment of the tricuspid chordae when necessary. Still other hearts can be corrected by re-routing the VSD to open into the pulmonary trunk, a manoeuvre which necessitates additional switching at either the arterial or atrial level [12]. In hearts with pulmonary or subpulmonary stenosis normally caused by a posterior deviation of the IS, however, the later procedure cannot be accomplished without leaving sub-aortic stenosis. The hearts with accompanying obstructive lesions initially in the subpulmonary area are those in which either the Rastelli or REV or Aortic Translocations procedures can be used [9-10-11].

**Surgical options**

There are varied surgical procedures reflecting the spectrum of conotruncal malformations. To understand the optimal applications is crucial to fully describe the anatomical interrelations of the various components of the ventricular outflow tracts. We sought to establish
Univentricular correction
(not included in this investigation).
Fontan type procedures are advocated when biventricular DORV repair is either impractical or extremely complex or at increased risk with a conventional biventricular repair including:

- Straddling Atrio Ventricular Valves.
- Atrioventricular Septal Defects.
- Hypoplastic Valve or Ventricle.
- Combination of Atrioventricular Septal Defects and Hypoplastic Valve or Ventricle.
- DORV with non committed or not-directly-committed VSD.
- DORV with intact VS.
Fig 9: In this case, the outlet septum is inserted across the doubly committed VSD, dividing it into subaortic (VSD1) and subpulmonary (VSD2) components. An apical continuation of the outlet septum forms a muscular shelf that sequesters the subpulmonary component of the morphologically right ventricle from the rest of the chamber, giving the spurious impression of complete transposition. In reality, the heart shows an extreme form of both double outlet and double-chambered right ventricle.

Fig 10: In this heart with double-outlet ventriculoarterial connection and doubly committed VSD, absence of the outlet septum together with absence of the septal components of both infundibulums permit the orifices of both arterial trunks to ride the crest of the ventricular septum. Fusion of the posterior limb of the septomarginal trabeculation (P) with the VIF forms a muscle bar separating the leaflets of the aortic and tricuspid valves.

**CONDUCTION TISSUES**  
*Fig. 11 (a, b), 12, 13, 14 (a, b), 15.*

**DOUBLE OUTLET RIGHT VENTRICLE WITH SUBAORTIC VENTRICAL SEPTAL DEFECT**

Fig 11: The differing infundibular morphology that is possible in DORV with subaortic ventricular septal defect (VSD). (a) (compare with Fig 5) The defect is perimembranous but there is a bilateral infundibulum. (b) (Compare with Fig 4) There is aortic-mitral-tricuspid continuity and still the defect is perimembranous. The anticipated site of the conduction tissue is superimposed on both photographs.
2. Material and methods

In this investigation we analyzed the anatomy of the Ventriculoarterial Connections following the sequential segmental basic concepts of an autopsied series of 33 selected hearts from the cardiopathological collection of the Royal Brompton Hospital with so called Cono Truncal Malformations and classified as having Fallot’s Tetralogy (5), Double Outlet Right Ventricle (23), or Complete Transposition with Ventricular Septal Defect (5). These data have been already presented in a previous specific publication [16]. A special concern is dedicated to the conduction tissue comparing the recent specimens to those presented in a investigation previously published on Surgical Anatomy of Double-Outlet Right Ventricle with Situs Solitus and Atrio Ventricular Concordance [15].

Morphology of the ventriculoseptal defect and conduction tissue

The perimembranous defects have in common the fact that an area of atrioventricular-aortic valvular fibrous continuity (central fibrous body) forms part of their rim. In all perimembranous defects, the atrioventricular node is confined within the triangle of Koch and the atrioventricular bundle penetrates at its apex. A long non branching bundle passes into the subaortic outflow in relation to the leading edge of the inlet septum. The abnormal position of the OS in these hearts needs to identify the defect as being perimembranous, muscular or subarterial. Then we complete the description of the VSD by describing into which portion of the right ventricular chamber the defect empties (inlet, trabecular, outlet portion or a combination), and also how the VSD relates to the arterial valves (subaortic, subpulmonary, non committed or doubly committed). The majority of the defects open into the outlet portion of the right ventricle. For morphological purposes the septal band of the TSM is a landmark in the right ventricle to differentiate the inlet from the outlet in different forms of DORV. The inlet VSD can be further associated with an atrioventricular septal defect. The term non-committed ventricular defect is not an anatomical term [19]. A committed VSD may open into the outlet portion of the right ventricle but the distance between the VSD and the semilunar orifice may be very extensive, either because of the extreme dextroposition of the aorta or a broad VIF and long OS. Moreover, the presence of aberrant chordae tendineae in the outlet of the right ventricle may disturb the direct relationship of the VSD to the semilunar orifice. In these circumstances it is more appropriate to define the VSD as not-directly-committed [20]. This VSD opens into the outlet portion of the right ventricle but is not directly subaortic or subpulmonary. The true non-committed VSD is located in the inlet portion.

Conduction tissue

- In DORV with subaortic VSD (Fig 11a, b-12).
- In DORV with subpulmonary VSD (Fig 13).
- In DORV with inlet muscular VSD (Fig 14a,b-15).

The conduction tissue is disposed as anticipated for the types of isolated perimembranous VSD. In all the perimembranous defects, the penetrating bundle passes through the central fibrous body in the posteroinferior rim of the defect branching on the left ventricular aspect of the septum. The non branching bundle is close to the edge of the defect in those hearts in which the defect extends into the inlet septum. In the subaortic and subpulmonary defects with muscular posterocaudal rims, the fusion of the PL of the TSM with the VIF protects the penetrating and non branching bundles from the edge of the defect. In the hearts with muscular inlet defects, the conduction tissue axis penetrates the ventricles cephalad to the defect.

Trabecula septomarginalis - outlet or infundibular septum and ventriculo infundibular fold (Fig 1a,b)

The TSM is a prominent trabecula plastered onto the right ventricular aspect of the septum which bifurcates in the outlet component of the ventricle into two limbs. The anterosuperior limb (AL) ascends.
DOUB-OUTF-DIT RIGHT VENTRICLE WITH SUBPULMONARY VSD
Fig 13: A muscular subpulmonary VSD with bilateral infundibulum. Note the insertion of the infundibular septum to the posterior limb of the TSM as compared with an insertion to the anterior limb when the defect is subaortic. (See Fig 12a and compare with Fig 7). The position of the conduction tissue has been superimposed.

to support the pulmonary valve. The posteroinferior limb (PL) overlays the inlet septum giving rise to the medial papillary muscle complex and other chordae to the septal leaflet of the tricuspid valve. Then the body of TSM becomes continuous with the major papillary muscles supporting the anterosuperior and inferior tricuspid valve leaflets. One band passes prominently across the cavity as the moderator band. The OS is below the distal part of the infundibulum and separates the ventricular outlets. The OS is small, merging with the upper frontally orientated part of the trabecular septum and inserts into the anterior septum to the AL of the TSM forming the inner component of the normal crista supraventricularis. Posteriorly the OS is not a septal structure separating the infundibulum from the outside of the heart. The outer component derived from the inner heart curvature is called the Ventriculoinfundibular Fold. This muscular structure separates an arterial valve from an atrioventricular valve and corresponds to the groove between the left ventricular sinus and the infundibulum. The OS may be displaced posteriorly into the left ventricular outflow tract producing subpulmonary stenosis in transposition of great arteries (Fig 2). The OS may be displaced anteriorly inserting to the AL of the TSM when the VSD is subaortic (Fig 3-4-5) or to the PL when the VSD is subpulmonary (Fig 6-7-8a,b). The OS may also be absent with doubly committed VSD (Fig 10). In Fig 9 the OS merges posteriorly with the VIF and the PL of the TSM. Inferiorly the OS divides the apical component of the right ventricle as an apical shelf of muscle giving the spurious impression of complete transposition. Double-outlet and double-chambered right ventricle.

3. Results

1. Tetralogy of fallot [13], (Fig 3-4)
The OS in TF is seen as an independent muscular structure and it is exclusively a right ventricular structure merging with the anterior limb of the TSM. The degree of overriding of the aorta depends on the extent of rightward displacement of the OS and the degree of formation of the VIF. The VSD is described as a malalignment and subaortic. The roof of the left ventricular margin of the defect is formed by the area of fibrous continuity between the leaflets of the aortic and mitral valves. The posteroinferior margin in most hearts is composed of fibrous continuity between the leaflets of the aortic mitral and tricuspid valves defining the defect perimembranous. When the posterior limb of the TSM fuses with the ventriculoinfundibular fold producing discontinuity between the leaflets of the aortic and tricuspid valves, the presence of this muscular tissue protects the conduction axis. There are hearts with close affinities to the Tetralogy, with fibrous continuity between the leaflets of the aortic and pulmonary valves (because the muscular outlet septum is completely lacking) with aortic overriding and stenosis of the ventriculopulmonary junction.

2. Double-outlet ventricle with subaortic ventricular septal defect [14-15-16], (Fig 5)
We define hearts as showing DORV when more than half of the leaflets of both arterial valves are supported by right ventricular structures. When defined in this fashion, the hearts can show the ventricularterial connection of double outlet together with the infundibular morphology of Tetralogy of Fallot. The outlet
DOUBLE-OUTLET RIGHT VENTRICLE WITH INLET MUSCULAR VSD

Fig 14 (a, b): The major differences in morphology between defects in the inlet muscular ventricular septum (a) when they are perimembranous and (b) when they have completely muscular remnants. (a) The position of the conduction tissues has been superimposed on both hearts, the position in (b) being shown as if viewed through the ventricular septum.

3. Double-outlet right ventricle with subpulmonary ventricular septal defect [17-18], (Fig 6,7)

When the OS is present but merges posteriorly with either the VIF fold or with the posterior limb of the TSM (or both), then the septal defect is subpulmonary. This arrangement usually produces a complete subaortic infundibulum anteriorly within the right ventricle the distal posterior of which can be freestanding sleeve of infundibular musculature.

Alternatively, the VIF can be attenuated to permit continuity between the aortic and tricuspid valvar leaflets. With this arrangement, when the pulmonary valve is positioned exclusively above the right ventricle, the ventriculoarterial connection is that of DORV. The presence or absence of fibrous continuity between leaflets of the pulmonary and mitral valves depends on the prominence of the VIF. A spectrum of malformation is seen in which the leaflets of the pulmonary valve come to be attached...
within the left ventricle. The overall spectrum of hearts with subpulmonary ventricular septal defect, can be considered to represent the Taussig-Bing malformation. Throughout this malformation, if the posterior limb of the TSM fuses with the VIF, there will be a muscular posteroinferior rim to the defect. If there is discontinuity between these structures, the leaflets of the mitral and tricuspid valves will be in fibrous continuity in the posteroinferior rim and the defect will be perimembranous.

4. Double outlet right ventricle with doubly committed VSD (Fig 10)
In these hearts there is complete absence not only of the muscular OS but also of the adjacent components of the infundibular musculature. The extent of the VIF determines the presence or absence of continuity between the leaflets of the arterial and atrioventricular valves. The relationship between the posterior limb of the TSM and the VIF determines whether the defect is perimembranous or has a muscular posteroinferior rim.

5. Transposition of the Great arteries with VSD [20], (Fig 8a,b-9)
When more than half of the circumference of the pulmonary valve is supported by the left ventricle, the ventriculoarterial connections are diagnosed as discordant rather than double-outlet. We may find anterior displacement of the infundibular septum and overriding pulmonary artery whereas posterior displacement has been reported to cause left ventricular outflow tract obstruction. In transposition of the great arteries without displacement of the infundibular septum, either arterial or atrial switch with transatrial closure of the VSD is applicable. Anterior displacement of the IS makes intraventricular rerouting from the left ventricle to the aorta difficult, therefore arterial switch with transatrial or transpulmonary closure of the defect without ventriculotomy is recommended. In hearts with posterior displacement of the IS, the anterosuperior rim of the defect is difficult to approach through the tricuspid valve and the tunnel from left ventricle to the aorta is straight. Hence the Rastelli or REV or Nikaidoh procedure is preferable. Depending on whether the VIF fuses or not with the posterior limb of the TSM, the VSD can be perimembranous or can have a muscular posteroinferior rim.

4. Comment
In this review, by a sequential-segmental descriptive approach we established the relations between Outlet Septum, Trabecula Septomarginalis, Ventriculo Infundibular Fold and Ventricular Septal Defect as hallmark of Conotruncal Malformations with Atioventricular concordance. We totally accept and promote the utility of a unified nomenclature avoiding artificial conventions and definitions but always outlining the anatomy to better understanding to which morphology each surgical principle can be better applied. We can summarize our findings as it follows:
- The Ventricular Septal Defect represents the only exit from the left ventricle and can be described combining the positional approach (subaortic, subpulmonary, doubly committed, non committed) with the morphologic concept of perimembranous, muscular and subarterial. Normally the defect is cradled between the limbs of the Trabecula Septomarginalis and the borders vary depending whether the Trabecula Septomarginalis fuses with the Ventriculoinfundibular Fold. It is essential to identify a defect as perimembranous or muscular so to predict the position of the conduction tissue. Our investigations has shown that it is the position of the Outlet Septum that determines the location of the Ventricular Septal Defect relative to the subaortic and subpulmonary outflow tracts.
- Other crucial anatomical aspects are the Outlet Septum, the Ventriculoinfundibular Fold and the Limbs (Antero Superior and Postero Inferior) of the Trabecula Septomarginalis. In the normal heart the left sided Ventriculoinfundibular Fold is absent and there is mitro-aortic continuity whereas the right sided Ventriculoinfundibular Fold as a fold of tissue separates tricuspid and pulmonary valves. In Conotruncal Malformations with subaortic Ventricular Septal Defect the Antero Superior Limb inserts into the outlet septum and the relationships of the Postero Inferior Limb with the Ventriculoinfundibular Fold determine mitro-aortic continuity or discontinuity. In Double-Outlet Right Ventricle with subpulmonary Ventricular Septal Defect the Postero Inferior Limb inserts into the Outlet Septum which may fuses with Ventriculoinfundibular Fold. When more than half of the pulmonary valve is supported by the left ventricle, the Ventriculo Arterial Connections are discordant and the Postero Inferior Limb inserts into the ventriculo infundibular Fold.
According to the spectrum of Cono Truncal Malformations there are many surgical options: with the iconographical documentation presented we have shown that the surgeon can have a clear understanding of the abnormal morphology based on a detailed analysis of the observed anatomy and this may be crucial in planning and applying different procedures. A standard classification may not be always possible and from the surgical stance is also not necessary.

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5. Bibliographie


Fig 1-10: Reprinted from «Annals of Thoracic Surgery»; Vol 59; Authors: Athos Capuani, Hideki Uemura,
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Fig 11 (a, b), 12, 13, 14 (a, b), 15: Reprinted from « Annals of Thoracic Surgery»; Vol 82 (3); Authors: Benson R. Wilcox, Siew Yen Ho, Fergus J. Macartney, Anton E. Becker, Leon M. Gerlis, Robert H. Anderson; Title: Surgical Anatomy of Double-Outlet Right Ventricle With Situs Solitus and Atrioventricular Concordance; Pages 405 - 417; Copyright 1981; with permission from « The American Association for Thoracic Surgery ».