An appreciation of the structural variability in the components of the ventricular outlets in congenitally malformed hearts

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Received 21 November 2006; received in revised form 10 January 2007; accepted 15 January 2007; Available online 23 February 2007

Summary

The morphology of the outlet septum, the ventriculo-infundibular fold, the infundibulum and the septomarginal trabeculation has been well described. These structures are closely interrelated, such that variations in the morphology and orientation of any one of them inevitably affect the morphology and orientation of the others. In this review, we seek to clarify how these normal relationships change within the setting of congenitally malformed hearts. This is of immense surgical significance, since the surgical correction of many congenital cardiac malformations involves procedures on, or in the vicinity of, these structures. We provide an insight into how the appearance of the components of the ventricular outlets can vary with the underlying pathology. We first define the relevant structures and describe how their location can vary in different malformations. We will then describe how the variations in that interrelationship may result in malalignment and deviation of the outlet septum, emphasising the surgical implications of these specific features.

Keywords: Outlet septum; Ventriculo-infundibular fold; Infundibulum; Septomarginal trabeculation; Septal band; Crista supraventricularis

1. Introduction

The outlet of the ventricular mass, supporting the origins of the arterial trunks, consists of the outlet septum (OS) separating the origins of one from the other, the ventriculo-infundibular fold (VIF) separating them from the atrioventricular valves, the potentially complete dual infundibulums, and is closely related to the septomarginal and septoparietal trabeculations found within the right ventricle. Their morphology is of immense surgical significance, since the surgical correction of many congenital cardiac malformations involves operating directly on, or in the vicinity of, these structures. We provide an insight into how the appearance of the components of the ventricular outlets can vary with the underlying pathology. We first define the relevant structures and describe how their location can vary in different malformations. We will then describe how the variations in that interrelationship may result in malalignment and deviation of the outlet septum, emphasising the surgical implications of these specific features.

2. Definitions (Fig. 1)

The ventriculo-infundibular fold is any muscle that is interposed between the leaflets of an atrioventricular and an arterial valve [1,2]. In other words, it is the muscle that separates the inlet of the ventricle from its outlet. When seen from the outside of the heart, this structure represents the inner heart curvature, which provides the support for the infundibular region of the ventricular mass. In the majority of hearts, however, at least in part, the muscular ventriculo-infundibular fold becomes attenuated, and is replaced by the fibrous tissue of atrioventricular—arterial valvar continuity. Whenever muscle is found between an atrioventricular and an arterial valve, however, it must be part of the ventriculo-infundibular fold. The key anatomical point is that the appearances of this structure are misleading. The fold can seem to represent a bulky mass of muscle when seen from inside the heart (Fig. 1). In reality, it is a relatively thin sheet of muscle that is folded back on itself (Fig. 2). Cutting through this muscular fold, of necessity, takes the surgeon outside the heart, and into the transverse pericardial sinus.

The infundibulum, or conus, is a complete muscular sleeve supporting the leaflets of one or the other, or sometimes both, of the arterial valves [3,4]. Such a complete muscular sleeve cannot exist when the relevant arterial valve is in fibrous continuity with its adjacent atrioventricular
valve, as this would be incompatible with complete muscular support of the leaflets of the arterial valve. Potentially, nonetheless, each of the two arterial valves can be supported by a separate and complete muscular infundibulum [2,5–10].

The outlet septum, or infundibular or conal septum, is any muscle that is interposed between the two ventricular outflow tracts. It separates the leaflets of the two arterial valves [6,11–13]. When present, it is usually muscular, but on occasion it can be represented by a fibrous remnant.

The septomarginal trabeculation (SMT), or septal band, is the extensive muscular strap that reinforces the right ventricular aspect of the ventricular septum, extending from the base to the apex of the morphologically right ventricle [14,15]. Towards the base, this muscular strap splits into the anterosuperior and the posteroinferior limbs. The former extends towards the outlet, and the latter towards the inlet of the ventricle. In the normal heart, these two limbs clasp the medial end of the ventriculo-infundibular fold, with a small amount of muscle at their junction representing the outlet septum. It is impossible in the normal heart, however, to recognise the part of the muscle that interposes between the outflow tracts. In malformed hearts, the components can spring apart, and in most instances the interventricular communication, if present, is then cradled between the two limbs of the septomarginal trabeculation. A series of muscle bundles radiates from the anterior margin of the septomarginal trabeculation towards the anterior free wall of the right ventricle. These muscle bundles are the septoparietal trabeculations [16], with the moderator band being the most prominent of these trabeculations.

3. Variations for each of the muscular structures

3.1. The ventriculo-infundibular fold

The ventriculo-infundibular fold may extend across the roofs of both ventricles, separating both atrioventricular valves from both outflow tracts. In most hearts, however, part of this muscular fold is effaced, such that one arterial valve is in fibrous continuity with its adjacent atrioventricular valve. This is seen in the left ventricle of most hearts, including the normal heart. The ventriculo-infundibular fold is then confined to the right ventricle (Fig. 1). Normal hearts, nonetheless, do exist with persistence of the fold in the roof of the left ventricle [17]. In other hearts, the fold may be quite small, and confined to the lateral parts of the right ventricle only, as in some examples of double outlet right ventricle (see below). In rare circumstances, the entirety of the fold may be attenuated. This is seen most frequently when both arterial trunks arise totally, or in their greater part, from the morphologically left ventricle.

3.2. The outlet septum

The outlet septum may be quite substantial, as in tetralogy of Fallot (Fig. 2), quite small and impossible to recognise, as in the normal heart, non-existent in common arterial trunk or replaced by a fibrous remnant in hearts with doubly committed juxta-arterial ventricular septal defects (VSDs) [18]. Its spatial orientation is also very variable, as

Fig. 1. Cartoon showing the nature and surgical significance of the outlet septum (OS), the ventriculo-infundibular fold (VIF) and the septomarginal trabeculation (SMT). The position of the first septal perforating artery (SPA) and the atrioventricular node and conduction axis (AVN) is marked. The major danger area is at the point of penetration of the conduction axis, albeit that this is protected when the posteroinferior limb of the SMT fuses with the VIF. The anterocephalad limb of the SMT can be excised to enlarge the ventricular septal defect (VSD), but there is the risk of damaging the SPA. It is safe to resect the VIF, albeit that this takes the surgeon outside the heart. The OS never contains the conduction axis, so it can be resected providing that care is taken not to damage the attachments of the arterial valves.

Fig. 2. Normal heart. The ventriculo-infundibular fold (VIF) is seen interposed between the inlet and the outlet of the right ventricle. The medial end of this fold is positioned between the two limbs of the septomarginal trabeculation (SMT). Inf: infundibulum, TV: tricuspid valve.
outlet right ventricle, or for both infundibulums to be deficient, as encountered in double outlet left ventricle [2,5,7,8,10]. In rare circumstances, a common infundibulum can support both arterial valves in the absence of a muscular outlet septum, as seen in the setting of the doubly committed and juxta-arterial ventricular septal defect, or rarely can support the solitary valve found in the setting of common arterial trunk.

3.4. The septomarginal and the septoparietal trabeculations

The main variation of surgical importance in the morphology of the septomarginal trabeculation is whether or not its posteroinferior limb reaches and fuses with the ventriculo-infundibular fold [19]. A ventricular septal defect is perimembranous when these two structures do not meet in the posteroinferior margin of the defect. In this setting, the atrioventricular conduction axis will be found penetrating the fibrous area of valvar continuity that forms the posteroinferior margin of the defect, and is at risk of being damaged during repair. When the posteroinferior limb of the septomarginal trabeculation does reach the ventriculo-infundibular fold, then the posteroinferior margin of the defect will be muscular (Figs. 4 and 6). This rim, depending on its size, provides protection to the conduction axis.

The main variation in the septoparietal trabeculations is whether or not they are hypertrophied [16]. It is this hypertrophy, when present, which provides the anterocephalad 'squeeze' seen in the subpulmonary infundibulum in the setting of tetralogy of Fallot.

4. The spatial relationship between the outlet septum and the ventriculo-infundibular fold

This varies in such a way that:

- the two structures are continuous;
- the two structures are distinct, but the outlet septum inserts onto the ventriculo-infundibular fold;
- the two structures are distinct and separate, the outlet septum merging with the septomarginal trabeculation independently of the ventriculo-infundibular fold;
- the outlet septum is absent;
- the ventriculo-infundibular fold is absent.

4.1. The two structures are continuous with each other

This is the commonest pattern, with the planes of the two structures coinciding. In this setting, as in the normal heart, it is necessary to rely on the positions of the atrioventricular valve and the two outflow tracts so as to infer the distinction of the outlet septum from the ventriculo-infundibular fold (Fig. 1). This arrangement is found in any heart with concordant ventriculoarterial connections and no override of either great artery, irrespective of the presence or absence of septal defects. Examples include hearts with a ventricular septal defect, those with pulmonary atresia with intact ventricular septum and those with atrioventricular septal defect and common atrioventricular junction. The pattern is also seen in heart with discordant ventriculoarterial connections, with or without a ventricular septal defect, but with no overriding arterial trunk.

The presence of an overriding arterial trunk typically results in greater prominence of the outlet septum, and a proportionately smaller ventriculo-infundibular fold. This is classically seen in tetralogy of Fallot (Fig. 2). When compared to the normal heart (Fig. 1), the overriding arterial valve can be seen to have moved towards the right, part of its origin being supported by right ventricular structures, and hence separated by the fold from the tricuspid valve. In this setting, the outlet septum, if present, becomes recognisable in its own right, while maintaining its continuity and alignment with the ventriculo-infundibular fold. In effect, in this setting, the override has resulted in the right ventricular component of the overriding arterial valve replacing the medial aspect of the ventriculo-infundibular fold. With greater degrees of override, the outlet septum becomes increasingly prominent, and the ventriculo-infundibular fold proportionately smaller. The relative extents of the ventriculo-infundibular fold and the outlet septum are then an additional variable in the morphological evaluation of these hearts.

4.2. The two structures are distinct, but the outlet septum inserts onto the ventriculo-infundibular fold

This arrangement is seen in hearts with double outlet right ventricle with side-by-side arterial trunks, particularly as in the so-called Taussig-Bing malformation (Fig. 3). In this setting, the ventriculo-infundibular fold, if complete, is interposed between the leaflets of the atrioventricular valves and both outflow tracts. The outlet septum, interposed between the two outflow tracts, extends from the
ventriculo-infundibular fold to the parietal free wall of the right ventricle. The precise angle of the junction between the outlet septum and the ventriculo-infundibular fold can vary depending on the spatial relationship between the two outflow tracts, but since it attaches to the fold rather than to the septomarginal trabeculation, the end result is that the interventricular communication opens beneath the left-sided arterial trunk, which is almost always the pulmonary trunk.

4.3. The two structures are distinct and separate

This is typically seen in hearts with double outlet right ventricle with anteroposterior arrangement of the outflow tracts (Fig. 4). Here, the ventriculo-infundibular fold is interposed between the atrioventricular valves and the outflow tracts, while the outlet septum is attached to the anterosuperior limb of the septomarginal trabeculation. This arrangement places the interventricular communication beneath the posteroinferior arterial trunk, which is almost always the aorta. This arrangement is also seen in our chosen example of tetralogy of Fallot with pulmonary atresia (Fig. 5).

4.4. The outlet septum is absent

This is seen in two settings. The first is in the setting of the doubly committed and juxta-arterial ventricular septal defect, where there is fibrous continuity between the two arterial valves on account of failure of muscularisation of the embryonic outlet septum, albeit that the outlet septum can be represented by a fibrous remnant. The second setting is in hearts with a common arterial trunk (‘truncus arteriosus’) (Fig. 6). In this setting, the outlet septum is never formed during cardiac development, leaving the heart with a common and solitary outflow tract.

4.5. The ventriculo-infundibular fold is absent

This very rare situation is typically seen in the setting of double outlet from the morphologically left ventricle, when there is fibrous continuity between the atrioventricular valves and both arterial valves.
5. Malalignment and deviation of the outlet septum

The spatial orientation of the outlet septum is determined by the relative positions of the two outflow tracts with respect to each other [5]. This can be anything within a spectrum of 360°, resulting in many different potential orientations and attachments of the outlet septum. The abnormally positioned outlet septum may be malaligned, or deviated. In some circumstances, the septum can be both malaligned and deviated. These two terms, therefore, must be distinguished from each other. As we have emphasised already, the outlet septum in the normal heart is a very small structure, found between the two limbs of the septomarginal trabeculation, where the ventriculo-infundibular fold joins the muscular ventricular septum. As we have also described, in the presence of override, the relationship of the outlet septum, if present, changes significantly, relative to the ventriculo-infundibular fold. It may be oriented in the same plane as the ventriculo-infundibular fold and remain continuous with it, join it at various angles that depend on the exact spatial interrelationship between the two outflow tracts, or be completely divorced from it. In all three variants, the outlet septum is no longer in the same plane as the muscular ventricular septum. This is referred to as ‘malalignment’. Indeed, the outlet septum can become exclusively a right, or exclusively a left ventricular structure.

In addition, the outlet septum may no longer insert between the two limbs of the septomarginal trabeculation. This provides it with the potential to become deviated either anteriorly or posteriorly, relative to the long axis of the outflow tract it separates. This is the essence of either anterocranial or posteroserial deviation, which, as indicated, usually coexists with malalignment.

6. Surgical implications

The recognition of the structures described above and their morphological variations guide the surgeon as to where sutures may be placed safely, and what may or may not safely be resected during the surgical procedures [20—22]. The outlet septum, in particular, can be an anchorage for suture lines during surgical repair. Safety, in this context, refers to the likelihood of damage to the conduction axis or to a major coronary artery, neither of which is found within the outlet septum.

The ventriculo-infundibular fold, irrespective of its extent and spatial orientation, is also devoid of conduction tissue, but it carries major coronary arteries within its epicardial aspect. As we have emphasised, although it looks like a thick muscle when viewed from inside the heart (Fig. 1), it is, in reality, a much thinner muscle that is folded back on itself (Figs. 2 and 7). Resection of this fold, therefore, takes the surgeon outside the heart to the atrioventricular grooves, which do contain major branches of the coronary arteries.

An infundibulum, if present, does not contain conduction tissue or major coronary arteries, although its base can be very close to the first septal perforating artery (SPA) [23,24]. Cuts through the walls of the infundibulum, of course, also take the surgeon outside the heart.

It is the various orientations and attachments of the outlet septum that primarily dictate which outflow tract is closest to the interventricular communication, and whether there is enough room to construct a baffle between the interventricular communication and the aortic outflow tract [21]. This information is indispensable when planning surgery, and is one of the most crucial components of the echocardiographic diagnosis.

The fusion of the posteroinferior limb of the septomarginal trabeculation with the ventriculo-infundibular fold in the presence of a ventricular septal defect determines whether the posteroinferior margin of the defect is muscular (Fig. 4), or whether the defect is perimembranous. This is surgically significant because, in the setting of a perimembranous defect, the atrioventricular conduction axis penetrates through the posteroinferior fibrous margin of the defect, and hence is at risk of being damaged during repair.

The anterior limb of the septomarginal trabeculation fuses with the outflow tract of the right ventricle. It carries a major septal perforating artery [23,24]. This is of particular concern in the Ross procedure, where the harvesting of the pulmonary autograft takes the surgeon dangerously close to this important blood vessel [4].

The septoparietal trabeculations are devoid of conduction tissue and major coronary arteries. They may safely be resected and sutured.

7. Comments

The morphology of the outlet septum, the ventriculo-infundibular fold, the free-standing infundibular sleeves and the septomarginal trabeculation are closely interrelated. Variations in the morphology and orientation of one of these structures inevitably affect the morphology and orientation of the others. The assessment of this interrelationship is vital in the understanding of the overall cardiac morphology and
diagnosis, as well as in the planning of surgical repair. The surgeon needs to recognise the variation in the extent of the outlet septum according to the underlying morphology. The surgeon must also appreciate the way in which the precise location of the outlet septum and its relationship to the other muscular components of the ventricular outlets vary in different malformations, particularly in terms of septal deviation and malalignment.

Despite the importance of the assessment of the above anatomical interrelationship, such evaluation should not, in any way, take precedence over rigorous initial sequential segmental analysis. Sequential segmental analysis remains the backbone of cardiac diagnosis. It starts by analysing each part of the heart independently of other parts, and ends by putting together all the gathered information, and cataloguing the additional defects, such as septal defects, patency of the arterial duct, anomalies of the aortic arch and so on. Assessment of the components of the ventricular outflow tracts, therefore, becomes relevant only towards the end of the sequential analysis. It is this assessment, nonetheless, that guides the surgeon in determining the best options for surgical repair of malpositioned outflow tracts, as well as pointing to those parts that can be removed without fear of damaging significant structures, such as the conduction tissues or the coronary arteries.

Acknowledgement

We are grateful to the department of histopathology at Birmingham Children’s Hospital for allowing us to examine and photograph some of the hearts from their collection for the purpose of this publication. Mrs Sarah Davies from that department was very helpful in arranging the sessions for the purpose of this publication. Mrs Sarah Davies from that department was very helpful in arranging the sessions for the purpose of this publication.

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