Review

The helical ventricular myocardial band of Torrent-Guasp: potential implications in congenital heart defects

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Summary

The new concepts of cardiac anatomy and physiology, based on the observations made by Francisco Torrent-Guasp’s discovery of the helical ventricular myocardial band, can be useful in the context of the surgical strategies currently used to manage patients with congenital heart defects. The potential impact of the Torrent-Guasp’s Heart on congenital heart defects have been analyzed in the following settings: ventriculo-arterial discordance (transposition of the great arteries), double (atrio-ventricular and ventriculo-arterial) discordance (congenitally corrected transposition of the great arteries), Ebstein’s anomaly, pulmonary valve regurgitation after repair of tetralogy of Fallot, Ross operation, and complex intra-ventricular malformations. The functional interaction of right and left ventricles occurs not only through their arrangements in series but also thanks to the structural spiral features. Changes in size and function of either ventricle may influence the performance of the other ventricle. The variety and complexity of congenital heart defects make the recognition of the relationship between form and function a vital component, especially when compared to acquired disease. The new concepts of cardiac anatomy and function proposed by Francisco Torrent-Guasp, based on his observations, should stimulate further investigations of alternative surgical strategies by individuals involved with the management of patients with congenital heart defects.

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1. Introduction

The new concepts of cardiac anatomy and physiology are based on the observations made by Francisco Torrent-Guasp’s discovery of the helical ventricular myocardial band [1—4]. When the established knowledge is challenged by new discoveries, a lot of time, discussions, and efforts are needed to accept new concepts, particularly when they are so revolutionary towards what has been taught over centuries [5—13]. The community of pediatric cardiologists and cardiac surgeons has to be involved in the evaluation of the new concepts of cardiac anatomy and physiology (the helical ventricular myocardial band described by Torrent Guasp) and their potential impact on the understanding and management of congenital heart defects.

The helical ventricular myocardial band will be analyzed in different settings:

- ventriculo-arterial discordance (transposition of the great arteries)
- double (atrio-ventricular and ventriculo-arterial) discordance (congenitally corrected transposition of the great arteries)
- Ebstein’s anomaly
- pulmonary valve regurgitation after repair of tetralogy of Fallot
- Ross operation
- complex intra-ventricular malformations

2. Embryology

Several studies have been performed on the embryology of the heart and great arteries, in order to investigate the development of the normal myocardial structure, as well as the occurrence of congenital heart defects [14—46]. While several studies have addressed the relationship between
embryology and the spiral arrangement of the myocardial fibers, very little has been reported about the potential impact of the observations made by Francisco Torrent-Guasp on the understanding of the heart development [20].

The developing human heart undergoes a transition with the following steps [19,23,26,29,33,35,37,38,40,41,43,44]:

a) a single vascular tubular structure (like in worms) in the 20-day embryo
b) a sequential pulsatile pump including an atrium, ventricle, bulbus cordis (conus), and truncus arteriosus (like in fish) in the 28-day embryo
c) a two-chamber heart with atrial and ventricular septal defects (like in amphibians and reptiles) in the 30-day embryo
d) a four-chamber structure (like in birds and mammals) in the 35–50-day embryo

The human heart seems to repeat within the first few weeks of the fetal life the evolution of the cardiac morphology which occurred in millions of years from worms to mammals. It has also been recognized that the myocardium of the embryologic tubular structure is not homogeneous, but partitioned into segments, defined as inflow tract, embryonic atria, atrio-ventricular canal, embryonic ventricles, and outflow tract [31,32]. To assist in the complicated twisting and repositioning of the segments, the primary heart tube must carry distinct axial information, in order to avoid any misstep in interpreting the left–right axial information [31,32]. Any disruption of the left–right axial information can result in a congenital heart defect, like persistent left superior vena cava, partial or total anomalous pulmonary or systemic venous connections, and atrio-visceral heterotaxy [17,31,32].

Over the last few years, many studies focused upon the identification of molecules showing asymmetric expression in the early embryo that are potentially involved in defining the left–right body plan, as well as observations on mutations in a gene responsible for left–right inversion, or situs inversus that result in the randomization of left—right orientation [16,17,19,21,22,25,28,31,32,35,36,38,43,46]. The developing heart loops, and the purpose of the heart looping is to convert the weak peristaltic force of a straight tube, as exists in worms, into a powerful pump as is found in fish: the greater the bend, the more powerful is the force [33]. By looping to the right, the heart is the first organ to break the bilateral symmetry of early development [17,31–33,35,36,44,47]. Since the dextro-ventricular loop is the first recognized sign of asymmetry and lateralization of the body organs, the clockwise spiralization of the heart should be considered a specific pattern of chirality in vertebrates.

The outflow tracts and the great arteries of the normal heart demonstrate a clockwise spiral pattern of development. The helicoid three-dimensional movement driving to this condition is genetically determined, appears in the dextro-ventricular loop (right ventricle to the right, left ventricle to the left), then progresses with further looping (anterior rotation of the right ventricle, posterior rotation of the left ventricle), and then finishes with the spiral septation of the outflow tracts and the great arteries [48,49]. Therefore, the spiral shape of the heart is the consequence of an asymmetric formation, and exists during coordinated three-dimensional developmental mechanisms involving the segments of ventricles and great arteries.

The genetically determined helicoid shape of the heart may be clockwise in patients with situs solitus, counter-clockwise in situs inversus, and such rotational changes may be either absent or interrupted in situs ambiguous [48]. The potential relationships between the spiral pattern of the heart and the segmental morphogenetic mechanisms were previously considered [48] in regard to the complex congenital heart defects with ventriculo-arterial discordance (transposition of the great arteries) and to the double discordance (congenitally corrected transposition of the great arteries) in our previous publication.

3. Ventriculo-arterial discordance (transposition of the great arteries)

The embryology leading to the congenital malformation with ventriculo-arterial discordance (transposition of the great arteries) has been quite extensively studied [14,19,34,48–52] related to the spiraling septation of the outflow tracts in the developing heart and is associated with the helical flow patterns.

In ventriculo-arterial discordance (complete transposition of the great arteries), the parallel relationship of the great arteries shows the interruption of the spiral pattern of the heart despite the presence of situs solitus of the atria and dextro-ventricular loop [48]. Ventriculo-arterial discordance might be included in the pathogenetic group of the defects of lateralization [48], as suggested by experimental animal research [53], clinical studies on familial recurrence [54], and analysis of human genetic mutations [55].

Supra-valvular pulmonary stenosis remains the most frequent medium and long-term late complications of the arterial switch operation, which is currently the accepted standard surgical treatment of children with transposition of the great arteries. Although several technical steps to reduce this problem, such as generous patching of the pulmonary sinus and extensive hilar dissection of the pulmonary arteries [56–59] were undertaken, there still remains a risk for this potential complication.

Focus upon the spiral relationship of the ventricular outflow tracts and of the great arteries in the normal heart makes it clear that the conventional arterial switch operation will result in complete loss of this relationship. For example, the anterior new pulmonary trunk (remaining at low pressure) is flattened when the Lecompte maneuver (mobilization of the pulmonary artery bifurcation anterior to the ascending aorta) is undertaken; this routine step in the arterial switch operation, may result in potential stenosis of the pulmonary artery branches with resultant increased peak velocity during systole. Failure to restore the high-pressure new ascending aorta into its natural location is a possible mechanism for this potential problem, so that the new pulmonary artery cross-sectional area becomes oval rather than remaining circular [60].

A ‘spiral arterial switch operation’ (Fig. 1) has been proposed [61,62] as a solution whereby reconnection of the great arteries to the appropriate ventricle may be done without losing the spiral relationship of the great arteries;
such a procedure re-creates the normal spiral relationship between the new aorta and the new pulmonary artery, as in normal hearts [61,62]. Support for this concept exists from computational fluid dynamic study analysis that compares the conventional arterial switch surgical technique (including the Lecompte maneuver) to the new technique of ‘spiral’ arterial switch; the spiral relationship has the functional superiority of more uniform velocity and wall-shear stress, and a smaller pressure drop and power loss ratio [63].

4. Double (atrio-ventricular and ventriculo-arterial) discordance (congenitally corrected transposition of the great arteries)

Previous studies of connections between the embryologic development and the malformation of double discordance [14,48,51] showed that the helicoids shape of the heart is inverted at ventricular level and interrupted at the level of the great arteries [48].

These patients present with atrial situs solitus, but with the simultaneous presence of levo-ventricular loop and transposition of the great arteries resulting in an anatomic pattern that is neither clockwise nor counterclockwise.

The potential implications of the anatomical observations by Torrent-Guasp on patients with double discordance must simultaneously consider (a) the associated heart defects (ventricular septal defect, pulmonary stenosis, malformations of the morphologically right atrio-ventricular valve on the systemic circulation) and (b) the available surgical options [64]. The double switch may be the best option because it restores morphologically left ventricle to the systemic circulation [64–67]. However, the spiral relationship of the two ventricular outflow tracts is not maintained by the arterial switch for transposition of the great arteries.

There is currently no evidence that the presence of a morphologically left ventricle in this malformation exists with the simultaneous capacity to have structure/function relationships similar to the normal left ventricle. This need sets the stage for further studies designed to search for the presence of a ventricular myocardial band in patients with double discordance, and to evaluate form/function relationship in the presence of inversion and interruption of the helicoids shape. We suspect that inversion and interruption of the helicoids shape accounts for the absence of a structurally normal ventricular myocardial band, and that this structural deficit is responsible for the observed impaired myocardial function. The observed suboptimal medium and long-term results of the double switch, despite technically adequate surgical procedures connecting the morphologically left ventricle to the systemic circulation, support this conclusion.

For these reasons the alternative option of ‘one and half ventricular type of repair’ (a procedure that includes end-to-side anastomosis of the superior vena cava to the right pulmonary artery in addition to the intra-cardiac repair, in order to reduce the volume overloading of the small/malfunctioning right ventricle) is now considered the
procedure of choice [64,68–71]. The surgical approach of one and half ventricular repair in double discordance treated by double switch procedure has several advantages. These benefits include (a) eliminating the risk of superior vena cava obstruction (present with conventional atrial rerouting); (b) leaving more intra-atrial space available for the pulmonary venous return; (c) volume unloading the right ventricle that is made smaller following right ventricle to pulmonary artery anastomosis of the superior vena cava to the right pulmonary artery. This procedure involves end-to-side implantation of the excision of the atrialized right ventricular wall with the cut edges sutured together. Reproduced with permission from Wu Q et al. Ann Thorac Surg 2004;77:470–6.

5. Ebstein’s anomaly

Ebstein’s anomaly involves anomalous development of the embryo that results in involvement of the right ventricular chamber and its atrio-ventricular valve [19,30,44]. Clinical signs closely relate to the degree of tricuspid valve regurgitation, right ventricular dysfunction, and associated lesions [56].

Surgical procedures for Ebstein’s anomaly include repair or replacement of the tricuspid valve and plication of the atrialized portion (Fig. 2) of the right ventricle [56]. Furthermore, a one-and-half ventricular repair is performed when there is reduced size and/or severely impaired function of the right ventricle. This procedure involves end-to-side anastomosis of the superior vena cava to the right pulmonary artery together with the intra-cardiac repair, in order to reduce the volume overload of the small/malfunctioning right ventricle [56,69,72,73].

Fig. 2. Right ventricular remodeling in Ebstein’s anomaly. Artist representation of the excision of the atrialized right ventricular wall with the cut edges sutured together. Reproduced with permission from Wu Q et al. Ann Thorac Surg 2004;77:470–6.

The occurrence of arrhythmias and impaired long term right ventricle function define the limitations of the conventional surgical techniques that reduce the right ventricular volume without restoring the normal right ventricular geometry, and have led to the development of a new procedure [74]. The components are shown in Fig. 3 where the atrialized portion of the right ventricle is not plicated, but rather completely excised in the shape of a trapezoid or triangle. The cut edges can be then sutured together to restore the right ventricular geometry and improve the relationship between structure and function [1–5]. The early results document a reduction of right ventricular cavity dimension, with a lesser incidence of right ventricular dysfunction and arrhythmia [74].

This example of improved right ventricular form and function relationships seems parallel to similar observations in adult patients that undergo left ventricular restoration.

Hopefully, development of better knowledge of the relationship between helical ventricular muscular band and right ventricular morphology in Ebstein’s anomaly may identify the optimum method of atrialized portion resection that can optimize right ventricular function.

6. Pulmonary valve regurgitation after repair of tetralogy of Fallot

Tetralogy of Fallot is a common cyanotic congenital heart defect that has undergone extensive studies of embryologic development [19,43,51]. Follow-up studies show that chronic pulmonary valve regurgitation after repair of tetralogy of Fallot often results in right ventricular dilatation and arrhythmias that produce late morbidity and mortality [56,68,75].

Factors responsible for the functional deterioration are:

a) right ventricular dilatation with unfavorable alteration in the right ventricular mass-to-volume ratio
b) scarring and fibrosis from the initial ventriculotomy, with late remodeling because of the pressure/volume overload
c) electrical uncoupling of ventricular contraction (wide QRS interval on the electrocardiogram) with right bundle branch block
d) left ventricular dysfunction

In the past, decision about pulmonary valve implantation alone, and its timing were considered the most important points determinant for recovery of the right ventricular function recovery in these patients [68,76]. Unfortunately, pulmonary valve implantation alone may not allow satisfactory reduction of right ventricular volume, or recovery normal systolic right ventricular function, when preoperative measurements document significant abnormalities of the end-diastolic volume >170 ml/m² or end-systolic volume >85 ml/m² [77] indices. These observations stress the importance of evaluating the importance of the right ventricular dilatation and its relationship with the right ventricular geometry, with particular emphasis on how changes in the structure may impact the helical ventricular myocardial band and its effect upon the right ventricular function.
These considerations led to surgical procedures that supplement pulmonary valve implantation with generous resection of the anterior wall of the right ventricle towards the apex, as a way to restore adequate right ventricular size and geometry (Fig. 3). Preliminary findings show that this valve and ventricle approach improves the right ventricular function as determined by contrast magnetic resonance imaging [78]. Subsequent considerations about right ventricular remodeling after treatment of pulmonary insufficiency after repair of tetralogy of Fallot, like with Ebstein's anomaly, may be enhanced by determining how the helical ventricular myocardial band described by Torrent-Guasp influences the surgical approaches to congenital heart defects.

7. Ross operation

The Ross operation consists in the transplantation of the native pulmonary valve in aortic position, with simultaneous replacement of the native pulmonary valve with a biological valved conduit [79,80]. The outflow tracts of both ventricles are involved in this surgical procedure, together with the inter-ventricular septum and its septal branch coronary arteries; the embryology of these areas were extensively studied [14,23,34,52,81], as well as the anatomy of the septal perforating arteries [82].

During the pulmonary autograft preparation for the Ross operation, it becomes evident that the pulmonary valve and the right ventricle constitute a separate entity attached simply through a thin layer of muscle. Donald Ross collaborated with Francisco Torrent-Guasp to define the interaction of the myocardial band during the pulmonary autograft procedure. Their collaborative dissections (Fig. 4) showed that the first septal branch of the left anterior coronary artery courses between the ascending and descending segments of the ventricular myocardial band [83]. Furthermore, surgical preparation of the pulmonary autograft must include separation of segments between the right basal segment and the ascending segment of the ventricular myocardial band (Fig. 5). To prevent subsequent pulmonary autograft regurgitation, a minimal amount of muscle should be removed [84], since the muscle is a not a supporting structure for the semilunar valve, as confirmed by the reported clinical results [80,85].
8. Complex intra-ventricular malformations

Further studies are required to determine the role of the helical ventricular myocardial band in a number of other complex intra-ventricular malformations that include double-inlet single ventricle, criss-cross heart, and certain types of double outlet right ventricle.

Reconstruction of the morphologic ventricle should include thoughts about the impact of restoring a more normal spatial configuration, as described in the helical ventricular band by Torrent-Guasp.

The ‘ideal’ surgical approach has not yet been defined. Decision options include whether the morphologically left ventricle is adequate to sustain the systemic circulation, or it be made to become with other available surgical approaches. This consideration is critical because bi-ventricular type may be extremely difficult, even if feasible in a ‘borderline left ventricle’ [86].

The elements for the decision-making process between uni- and bi-ventricular type of repair include morphometric and functional parameters, hemodynamic data, available surgical options, results of the personal and institutional experience. However, we do not now have a satisfactory answer to the question ‘Is a high-risk bi-ventricular repair always preferable to conversion to a single ventricle repair?’ [87]. This dilemma opens the door to questioning if concepts about the helical ventricular myocardial band can help us to understand the relationship between form and function in uni-ventricular hearts. Partial or complete absence of the inter-ventricular septum in this malformation leads to absent spiral motion of this critical structure, and may be the vital factor responsible for the poor cardiac function. Our knowledge about matching form and function shall grow, since vast improvements have occurred in non-invasive studies in human beings and anatomical studies in animal species with congenital heart defects. This new knowledge shall help in the further evaluation of the helical ventricular myocardial band in the various congenital heart malformations.

9. Conclusions

The functional interaction of right and left ventricles occurs not only through their arrangements in series but because of common structural spiral features [1–5,13]. Changes in size and function of either ventricle may influence the performance of the other ventricle, and this is particularly evident in the neonatal myocardium [88].

The variety and complexity of congenital heart defects makes understanding the form/function relationship more complex than currently exists for acquired heart disease.

Nevertheless, the new concepts of cardiac anatomy and function, based on the observations by Francisco Torrent-Guasp, should stimulate the individuals involved with the management of patients with congenital heart defects to consider further investigations in order to expand their horizons and use this information to further consider and evaluate alternative surgical strategies.

References


