Surgical anatomy of the coronary circulation in hearts with discordant atrioventricular connections

Abstract We examined the arrangement of the coronary arterial and cardiac venous systems in 46 specimens with discordant atrioventricular connections so as to identify any structural abnormalities and to consider their surgical implications in terms of anatomical biventricular repair. Grossly abnormal arterial courses were seen in 11 hearts (24%). A substantial branch supplying the morphologically right ventricular outflow tract, which could restrict a ventriculotomy, was found in 61% of cases. The coronary sinus received all the morphologically right ventricular veins, as well as the posterior interventricular vein, in 40 hearts, this pattern being in contrast to the pattern in the normal heart. The morphologically left ventricular and anterior interventricular veins, all of which drain via the coronary sinus in the normal heart, were frequently connected independently to the morphologically right atrium in the specimens with discordant connections, the drainage occurring through the spaces between the pectinate muscles. These direct drainages are at risk of potential damage either by extensive intra-atrial maneuvers or by postoperative intra-atrial thrombosis. It is predicted, therefore, that surgical results can be improved still further when account is taken of this vascular anatomy of the heart itself.

Key words Congenitally corrected transposition • Discordant atrioventricular connections • Coronary artery • Cardiac vein • Double switch operation

Introduction

The functional unsuitability of the morphologically right ventricle as a systemic ventricle has long been stressed in the setting of congenitally corrected transposition (discordant atrioventricular and discordant ventriculo-arterial connections) [10, 16–18, 20, 28], although the topic remains controversial [4, 7]. In order to achieve better surgical outcomes in the long term, trials of anatomical repairs by intraventricular rerouting or the arterial switch procedure concomitant with intra-atrial rerouting are increasingly used for definitive repair [6, 12, 13, 29]. As has previously been reported in our series of such operations performed in Osaka [29], we have experienced a coronary arterial problem related to one of the deaths in the medium term. In this particular patient, a substantial infundibular branch originating from the left-sided morphologically right coronary artery crossed the right ventricular outflow tract, which was subaortic. The course of this artery, in association with that of the anterior interventricular artery, considerably restricted the ventricular incision for enlargement of the stenotic right ventricular outflow tract using a patch. The obstruction of the anterior interventricular artery, which progressed after the operation, was probably caused by the surgical maneuver close to the course of the major artery. This experience convinced us that detailed morphologic information and a better understanding of the
Coronary arteries can point the way to improving overall surgical results and establishing the optimal strategy. With this in mind, we examined the anatomy of the coronary arteries and the cardiac veins in autopsied specimens with discordant atrioventricular connections.

Materials and methods

Forty-six hearts with discordant atrioventricular connections were examined at the National Heart & Lung Institute in London (25 hearts), the Children's Hospital in Pittsburgh (15 hearts) and at the National Cardiovascular Center in Osaka (6 hearts). These specimens represent all the autopsied hearts with this peculiar segmental combination stored at these three institutions in which it proved possible reliably to identify the arrangement of the coronary arteries and the cardiac veins. The usual atrial arrangement was seen in 42 hearts, with mirror image arrangement in 4. Ventriculo-arterial connections were discordant in 21, double outlet from the morphologically right coronary artery in 13, and single outlet via the aorta from the right ventricle in 12. The aorta was left-sided and anterior to the pulmonary trunk in all with the usual atrial arrangement except for one heart in which the aorta was right-sided and anterior. Such a right-sided and anterior location of the aorta relative to the pulmonary trunk was the rule in the four cases with mirror-imaged arrangement of the atria. All the coronary arteries and cardiac veins were carefully dissected and macroscopically inspected to determine their courses on the surface of the heart mass and their orifices within the aortic or atrial walls.

In order to describe the orifices of coronary arterial origins from the aortic sinuses, the terms "left-hand facing sinus" and "right-hand facing sinus" were used as perceived from the stance of the observer located in the pulmonary trunk and looking toward the aorta (Fig. 1) [1, 24]. As for epicardial courses, major branches were described in accordance with their locations on the ventricular mass such as "the morphologically right/left ventricular arteries" and "the anterior/posterior interventricular artery." These were described as perceived from the standpoint of the observer located in the pulmonary trunk and looking toward the aorta. Epicardial courses of major coronary arteries were described as "anterior/posterior interventricular artery" and "morphologically right/left ventricular arteries", in combination with "morphologically right coronary artery" and "morphologically left circumflex artery" comparable to those in normally structured heart [1, 9, 21, 22] (Fig. 1, Fig. 4), although the arterial origins from the aorta

Results

Coronary arteries

Out of 46 hearts, 35 (76%) had the coronary arterial pattern comparable to that seen as the commonest variant in complete transposition (concordant atrioventricular and discordant ventriculo-arterial connections) [1, 9, 21, 22] (Fig. 1, Fig. 4), although the arterial origins from the aorta...
Fig. 2 Cardiac venous system in normal heart and the most common pattern in corrected transposition. A drawing modified from that in the article by Walsmley, 1929 (viewed from behind). The coronary sinus was found at atrioventricular groove between morphologically left atrium and morphologically right ventricle, in any case with discordant atrioventricular connections.

were in a reversed pattern in which the left-sided morphologically right coronary artery originated from the left-hand facing sinus (sinus #1) and the right-hand facing sinus (sinus #2) gave rise to the main stem of the anterior interventricular artery and the morphologically left circumflex artery in the setting of the usual atrial arrangement. Abnormal origins and courses were seen in the other 11 hearts. All branches arose from a single main stem in four cases, the main stem originating from the left-hand or the right-hand facing sinus in two each. Another two hearts possessed an isolated anterior interventricular artery along with an abnormal proximal course of right-sided morphologically left ventricular artery (morphologically left circumflex artery) posterior to the pulmonary trunk. Dual anterior interventricular arteries, one of which originated independently, were found in one. Abnormal branching of part of the morphologically right coronary artery was seen in three examples, either having independent origin from the aorta (in two) or arising from the anterior interventricular artery (in one). In the remaining heart, both orifices for the morphologically left and right coronary arteries were identified within the left-hand facing sinus, the common stem for the anterior interventricular and the left ventricular arteries coursing posteriorly to the atretic pulmonary trunk.

A main coronary arterial branch coursing anterior to the pulmonary trunk was found in 44 hearts (96%). This branch arose from the anterior interventricular artery (in 12), from the morphologically right coronary artery (in 12), or from a right ventricular artery (in 4). Such an additional branch was seen more frequently in hearts with double outlet from the morphologically right ventricle than in others ($P=0.016$ by chi-square test).

In terms of dominance of coronary arterial perfusion for the posterior wall, 27 hearts (59%) had one or more morphologically left ventricular arteries supplied by the morphologically right coronary artery. In another 19 hearts, the morphologically right coronary artery did not go far beyond the crux of the heart, but terminated as the posterior interventricular artery.

Cardiac veins

The coronary sinus, as defined by the cited anatomical features, was recognized in all cases we investigated. It was always located on the side of the morphologically left atrium. Its opening was recognized within the morphologically right atrium in all hearts but two. In the exceptional cases, the orifice of the coronary sinus was found within the morphologically left atrium in association with an intramural course of the venous lumen.

The anterior interventricular vein drained directly into the appendage of the morphologically right atrium in 40 specimens (87%), or was connected to a right-sided circumflex vein with (in 1) or without (in 3) drainage via the
All of the veins draining directly into the atria opened through orifices adjacent to the atrioventricular groove and within the portion of the atrial wall covered with pectinate muscles.

**Discussion**

The anatomical disposition of the atroioventricular conduction tissues has been justifiably emphasized in hearts with discordant atrioventricular and ventriculo-arterial connections so as to avoid surgical damage at the time of closure of ventricular septal defects [3,8,14,23,25]. With the increasing use of extensive surgical techniques in recent years [6,12,13,29], other morphological features should now be taken into account if results are to continue to improve. In terms of principles, the so-called double switch operation consists of an atrial baffling procedure combined with either an arterial switch operation or an intraventricular rerouting procedure. For successful arterial switching, the anatomy of the coronary arteries, particularly their aortic origin, is of obvious surgical importance. For effective intraventricular rerouting concomitant with reconstruction of the pathway from the morphologically right ventricle to the pulmonary arteries, it will also be helpful to be fully acquainted with any coronary arterial courses which might restrict incisions to the morphologically right ventricle. When employing intra-atrial maneuvers for atrial redirection, however, the anatomy of cardiac veins should also be examined carefully. The purpose of this study, therefore, was to provide a morphological understanding of the overall coronary circulation.

As concerns the coronary arterial orifices and courses, our result was consistent with the previous descriptions [2,15]. The commonest anomaly was the solitary coronary arterial system seen in 4 of 11 hearts with abnormal origins and courses, the same comment having been made in one of these previous articles [2]. The finding of a larger arterial branch crossing the morphologically right ventricular outflow tract in 61% of hearts is of technical importance for effective achievement of intraventricular rerouting and reconstruction of the pathway from the right ventricle to the pulmonary arteries. An appropriate site for a ventriculotomy should be selected after careful observation of such branches. This is particularly crucial in hearts with double outlet or single outlet from the right ventricle, since in these hearts intraventricular rerouting is often a surgical option of choice because of the coexisting pulmonary stenosis or atresia. Although a confident opinion may exist that such infundibular (conal) branches can be sacrificed without any problems, these branches seemed too substantial to be imprudently cut and ligated when aiming for successful achievement of the extensive anatomical repair. The region anterior to the connection between the ventricle and the pulmonary trunk is unlikely to be suitable for a surgical in-

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**Fig. 3** Venous valves between coronary sinus and other cardiac veins. The coronary sinus was opened and viewed from behind. The valve of Veuvesens and other venous valves are demonstrated in a heart with discordant atrioventricular connections.
Fig. 4 Orifices and courses of the major coronary arteries. All major patterns are shown in light of the variety of ventriculo-arterial connections. Incidence of significantly abnormal branching was 25%. Relatively substantial artery supplying outflow tract of morphologically right ventricle was more common in hearts with double outlet right ventricle than in others ($P = 0.016$, chi-square test). Patterns in 4 hearts with mirror-imaged atrial arrangement were described by mirror image projection. VA, ventriculo-arterial; mRV, morphologically right ventricle.

Coronary arteries

<table>
<thead>
<tr>
<th>discordant VA connections (without pulmonary stenosis)</th>
<th>Aorta arising from mRV &amp; pulmonary stenosis</th>
<th>double outlet mRV (without pulmonary stenosis)</th>
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<tr>
<td><img src="image1.png" alt="Diagram 1" /></td>
<td><img src="image2.png" alt="Diagram 2" /></td>
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number of hearts, (*) : hearts with mirror-imaged arrangement

Fig. 5 Connections of longitudinal veins. Drainage patterns of cardiac veins were frequently unusual, the anterior interventricular vein and some of morphologically left ventricular veins being directly connected to morphologically right atrium. In normal heart, these veins almost always drain via the coronary sinus. Patterns in 4 hearts with mirror-imaged atrial arrangement were described by mirror image projection. mRA, morphologically right atrium; mLV, morphologically left ventricle; mLA, morphologically left atrium.

Cardiac veins

<table>
<thead>
<tr>
<th>anterior interventricular vein</th>
<th>posterior interventricular vein</th>
<th>morphologically left ventricular veins</th>
<th>morphologically right ventricular veins</th>
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<td><img src="image4.png" alt="Diagram 4" /></td>
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number of hearts, (*) : hearts with mirror-imaged arrangement
cission. In addition to potential damage to the conduction tissues, distortion or injury of the major coronary arteries is very likely to occur.

The cardiac venous system remains poorly studied in the majority of malformed hearts. To the best of our knowledge, this is the first study to provide morphological information regarding the cardiac venous system in the setting of discordant atrioventricular connections. In the normal heart, all the left ventricular veins, as well as the anterior and posterior interventricular veins, drain into the right atrium via the coronary sinus. The coronary sinus, therefore, is normally a component of the vestibule of the morphologically left atrium, and drains the blood from all of the morphologically left ventricle and a significant part of the morphologically right ventricle. Only a small proportion of the right ventricular veins drain directly into the right atrium, these being the ones returning the blood from the anterior wall of the right ventricle and opening directly through orifices between the pectinate muscles into the floor of the right atrial appendages (the short cardiac veins) [11, 26] (Fig. 2). Compared with these anatomical features of the normal heart, the ventricular parts draining via the coronary sinus were markedly different in the setting of discordant atrioventricular connections.

This is no more than to be expected since, as a consequence of the discordant atrioventricular connections, there is disharmony between the components of the atrial and ventricular chambers. Thus, the coronary sinus, a component of the morphologically left atrium, drains blood predominantly from the morphologically right ventricle. The anterior interventricular vein, which is a component of the great cardiac vein in the normal heart, is no longer located in such a position that it can drain to the coronary sinus unless it takes a circumflex course round the junction between the morphologically right atrium and the morphologically left ventricle. Only in 15% of the hearts studied was all the venous blood drained from the morphologically left ventricle to the coronary sinus via such circumflex veins in the morphologically right atrioventricular junction. Instead, many more veins drained directly to the pectinated floor of the morphologically right atrium in the fashion of the short cardiac veins of the normal heart. In the preoperative state, these differences may not be significant, because the morphologically right ventricle is the systemic ventricle in hearts with discordant atrioventricular connections, and the venous cardiac blood returning from this systemic ventricle continues to be drained via the coronary sinus. The differences, nonetheless, may be of much more significance during and after corrective surgical procedures.

Thus, the method of retrograde cardioplegia cannot be appropriate for hearts with discordant atrioventricular connections. If the technique should be used, a considerable part of the musculature of the morphologically left ventricle would not be, or would be insufficiently, perfused by the cardioplegic solution as seen in the morphologically right ventricle in the normal heart [19], and could suffer damage. The damage would be enhanced when topical cooling of the left ventricular mass is incomplete by virtue of its anterior location. Furthermore, the presence of relatively competent venous valves adjacent to the coronary sinus also may mitigate against effective perfusion (Fig. 3) [5]. Accordingly, our recommendation is for antegrade infusion of cardioplegic solutions in this setting.

Surgical injury to the venous orifices could also be of significance. The direct connections of longitudinal veins to the atrium necessarily possess openings through an atrial wall covered with pectinate muscles, and the orifices are virtually impossible to be seen clearly. Such independent openings of the longitudinal veins to the atrium, therefore, can either be injured directly at the time of extensive intraatrial maneuvers, or occluded postoperatively by intraatrial formation of thrombus or peel. It remains unclear whether such impaired venous drainage would produce any problem in the clinical course. The capillary network within the myocardium may provide an alternative venous pathway if one or some of the cardiac veins are occluded. Since there is still no unequivocal evidence, however, that the alternative pattern of venous drainage is perfect for the entire coronary circulation, it seems sensible, to us, to avoid surgical damage to the cardiac venous system as much as possible. Insufficient coronary circulation, if it occurs in the early postoperative stage after extensive surgery, could enhance the myocardial edema which has already been evoked by the use of cardiopulmonary bypass and cardiac arrest, and might produce risks militating against successful surgery, intra-atrial maneuvers should avoid the atrial pectinate muscles close to the atrioventricular grooves, and postoperative administration of anticoagulants should be thoughtfully planned. In addition, it is a fact that a considerable proportion of the venous drainage from the morphologically left ventricle will drain, after the so-called double switch procedure, into the systemic atrial chamber rather than into the atrial cavity for the pulmonary circulation, thus leaving a minimal right-to-left shunt.

From the diagnostic stance, it remains to be established whether these unusual patterns of the cardiac veins can be precisely diagnosed prior to surgery. It would be particularly difficult to identify the entire system of cardiac veins. We emphasize, therefore, that the preoperative appreciation of the possible abnormalities shown in this study could be of clinical help. As for the coronary arterial variations, the precise evaluation prior to anatomic biventricular repair, by either angiography, echocardiography, or inspection during the operation, are of surgical importance. We conclude that, with the morphological findings in the coronary arterial and venous anatomy presently described, there is still room for improvements in surgical strategies and techniques for patients with discordant atrioventricular connections.
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