CONGENITAL HEART DISEASE

Hearts with concordant ventriculoarterial connections but parallel arterial trunks

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Objectives: To determine the characteristic morphological features of hearts with concordant ventriculoarterial connections and parallel arterial trunks, and to provide unequivocally a method to describe their anatomy.

Design, methods and patients: The entire cardiac database and cardiac pathological archive at the Hospital for Sick Children, Toronto, Ontario, Canada, was interrogated to identify all patients with concordant ventriculoarterial connections and parallel arterial trunks. The clinical records, autopsy reports and actual cardiac specimens of those who underwent autopsy, were reviewed.

Results: 8 cases meeting our criteria were identified. The infundibular anatomy was variable, including four hearts with bilateral infundibulums, three with subpulmonary infundibulums and one with bilaterally absent infundibulums. Considerable variability was also found in the type of atrial arrangement, along with the morphology of the atrioventricular junctions. The most common findings were the usual atrial arrangement (n=5), left juxtaposition of the right atrial appendages (n=3), an atrial septal defect (n=6), univentricular atrioventricular connection (n=5), ventricular septal defect (n=8) and pulmonary obstruction (n=4). In addition, five specimens had either a single coronary artery or two coronary arteries arising from the anticipated right coronary aortic sinus.

Conclusions: Concordant ventriculoarterial connections with parallel arterial trunks can be found in a variety of segmental combinations. An accurate diagnosis of these rare hearts can be achieved by detailed analysis of not only the ventriculoarterial connections but also the infundibular anatomy and the spatial relationship of the arterial trunks. Particular attention to the coronary arteries is warranted.

Major advances in diagnostic techniques have meant that most features of the abnormal heart can now be identified with comparable precision during life as in the autopsy room. One group of lesions that still gives problems in understanding is the so-called anatomically corrected malposition, and in particular, its relationship to the other esoteric anomalies called isolated ventricular inversion and isolated ventricular non-inversion. For many years, these lesions were considered to be embryologic impossibilities.¹ Detailed analysis of the specimens in pathological archives proved their existence,^{2–4} and subsequently they have been recognised during life.^{5–7} The precise features permitting diagnosis remain controversial.⁸ In this article, we describe these features and show how separate attention to the fashion in which the arterial trunks arise from the ventricles, as opposed to infundibular morphology and arterial relationships, clarifies any persisting disagreements about their anatomy.

DESIGN, SETTING AND PATIENTS

We searched the cardiac database and cardiac pathological archives of the Hospital for Sick Children, Toronto, Ontario, Canada, to identify all patients recognised with the arterial trunks arising from morphologically appropriate ventricles, but with parallel rather than spiralling arterial trunks. We reviewed the clinical history, investigations and available autopsy reports of those who had undergone necropsy. In addition, we undertook a detailed review of the cardiac specimens, analysing all hearts to account for all cardiac segments and junctions, with particular attention to the ventriculoarterial connections, infundibular morphology and the spatial relationship between the intrapericardial arterial trunks.

RESULTS

We identified a total of eight heart specimens available for review. These patients were born between 1964 and 1973.

Case 1

This infant had cyanosis immediately after birth. Soon thereafter, congestive heart failure ensued. The clinical impression was that of transposition with ventricular septal defect. She failed to thrive, and the problems were difficult to control medically. Catheterisation at age 3 months suggested either transposition with a large ventricular septal defect or double outlet right ventricle of the Taussig–Bing type. Banding of the pulmonary trunk was undertaken, but the infant died of early postoperative respiratory complications.

Examination of the heart showed the usual atrial arrangement with left-sided cardiac apex. The venoatrial connections were as expected, and the atrial septum was intact. The atrioventricular connections were discordant, with left-handed ventricular topology (fig 1A,C). Both ventricles were complete, with the atrioventricular junctions guarded by morphologically tricuspid and mitral valves, respectively, which were of good size. The ventriculoarterial connections were concordant, with the aortic root arising from the right-sided morphologically left ventricle, with fibrous continuity between the leaflets of the mitral and aortic valves (fig 1B). The pulmonary trunk was supported above the left-sided morphologically right ventricle by a complete muscular infundibulum (fig 1C). The arterial trunks exited from the base of the heart in parallel fashion, with the aorta located rightward and slightly posterior relative to the pulmonary trunk (fig 1B). The aortic valve, ascending aorta and the transverse aortic arch, which was left-sided, were hypoplastic. Associated anomalies included a perimembranous

Figure 1 Autopsy findings in case 1. (A) The right atrium (RA) joins the morphologically left ventricle (MLV) through a morphologically mitral valve. By reflecting the mitral valve (B) the aorta (AO), originating from the right-sided morphologically left ventricle, and the fibrous continuity between the leaflets of the aortic and mitral valves (arrow) are shown. The parallel arrangement of the aorta and pulmonary trunk (PT) is evident. The left side of the heart (C) shows the left atrium (LA) communicating with the left-sided morphologically right ventricle (MRV) through the tricuspid valve. The arrow shows the completely muscular subpulmonary infundibulum.

ventricular septal defect and a solitary coronary artery arising from the right coronary aortic sinus.

Case 2

This child was suspected of having congenital cardiac disease since early infancy, and experienced frequent respiratory infections and problems with feeding. She was referred to the Department of Pediatrics, Hospital for Sick Children, Toronto, Ontario, Canada, at age 22 months. After investigations, the presumptive diagnosis was congenitally corrected transposition with ventricular septal defect and cleft mitral valve. Surgery was scheduled to close the ventricular septal defect and repair the mitral valve, but was not completed because of the operative finding of a functionally single ventricle. The child died on the first postoperative day due to respiratory complications.

Examination of the cardiac specimen showed the usual atrial arrangement, with normally positioned atrial appendages and normal systemic and pulmonary venous connections. The atrial septum was intact. The atrioventricular connections were concordant (fig 2A), and right-handed ventricular topology was seen. The mitral valve and left ventricle were hypoplastic. The ventriculoarterial connections were concordant, with bilateral infundibulums (fig 2B,C). The arterial trunks exited from the ventricular base in a parallel fashion (fig 2D). The coronary arterial pattern was normal. A large ventricular septal defect was present.

Case 3

This newborn was admitted with worsening cyanosis. Assessment suggested mirror image arrangement with a right-sided heart, with tricuspid atresia and pulmonary stenosis. The child died soon after admission.

Examination of the heart showed that the ventricular apex pointed to the right, and there was isomerism of the morphologically right atrial appendages, with left-sided juxtaposition of the right-sided appendage (fig 3A). Bilateral superior caval veins, each opening to the atrial roof adjacent to a terminal crest, and a bridging brachiocephalic vein were seen. The inferior caval vein drained to the right-sided atrium, and the coronary sinus was absent. The pulmonary veins all drained anomalously to the left superior caval vein. The atrioventricular connections were biventricular, but guarded

by a common atrioventricular valve (fig 3B). A right-handed ventricular topology was seen, with a hypoplastic right ventricle. A typical ventricular component of an atrioventricular septal defect was present between the ventricular inlets. The ventriculoarterial connections were concordant, with the aorta exiting from the heart in a parallel fashion, but anterior and to the left of the pulmonary trunk. Both arterial valves were supported by complete infundibulums (fig 3C,D), with subvalvar and valvar pulmonary stenosis (fig 3D). A solitary coronary artery arose from the right coronary aortic sinus.

Case 4

This infant was admitted at age 8 days because of difficulties in feeding and cyanotic spells. Catheterisation at age 6 months showed the heart to be right-sided with the usual atrial arrangement, but left-hand ventricular topology was observed in the setting of a ventricular septal defect and pulmonary stenosis. A Waterston shunt was performed, followed by a right Blalock–Taussig shunt and pulmonary valvotomy. The child died at 6 months from sepsis after surgery.

Examination of the specimen showed the usual atrial arrangement, with left juxtaposition of the right atrial appendage and the cardiac apex pointing to the right. The venoatrial connections were physiologically normal, albeit that a left superior caval vein drained into the right atrium through the coronary sinus. A large defect was found in the floor of the oval fossa. The right atrioventricular connection was completely absent, with the left atrium connected to a dominant morphologically left ventricle through a widely patent mitral valve (fig 4A). The incomplete morphologically right ventricle was right sided and anterior, being fed from the dominant ventricle through a muscular ventricular septal defect (fig 4B). The ventriculoarterial connections were concordant. The aortic valve, supported exclusively within the dominant ventricle, was in fibrous continuity with the mitral valve (fig 4A). The aorta exited from the heart parallel to the pulmonary trunk, which was supported above the right ventricle by a complete infundibulum. Subvalvar and valvar pulmonary obstructions (fig 4B) were seen. The arterial trunks ascended from the heart in parallel fashion, with the aorta anterior and to the left. The aortic arch was left sided and unobstructed. A single coronary artery arose from the right coronary aortic sinus (fig 4A).

Figure 2 Autopsy findings in case 2. The atrioventricular connections were concordant. (A) The right atrium (RA) connects to the morphologically right ventricle (MRV). (B) The pulmonary trunk (PT) arises from the right ventricle, with a complete subpulmonary infundibulum (dotted line), and the aorta (AO) arises from the left ventricle in the roof of the ventricular septal defect (VSD). (C) The complete subaortic infundibulum (dotted line) interposes between the leaflets of the aortic and mitral valves in the roof of the left ventricle (MLV). (D) The parallel arrangement of both arterial trunks.

Case 5

This girl had cyanosis at birth, and failed to thrive in early infancy. She underwent a first cardiac catheterisation at age 4 months, which suggested transposition with ventricular septal defect. At age 5 years, a second catheterisation suggested double-inlet ventricle, discordant ventriculoarterial connections and infundibular stenosis. A classic Glenn operation was performed, along with banding of the pulmonary trunk. At age 11 years, a third catheterisation showed the usual atrial arrangement, double-inlet left ventricle, hypoplastic left-sided morphologically right ventricle and concordant ventriculoarterial connections, albeit with the aorta exiting from the heart parallel to the pulmonary trunk. The pulmonary trunk, by now, was aneurysmal because of the central shunt, and there was

severe pulmonary hypertension. The patient had seizures and a cardiac arrest 1 h after catheterisation, from which she did not recover.

Examination of the specimen showed the usual atrial arrangement with left-sided cardiac apex. The systemic venous connections were normal, but there was totally anomalous pulmonary venous return to the coronary sinus. A defect was observed in the floor of the oval fossa. Both atriums drained to the dominant left ventricle through separate right and left atrioventricular valves, and the left ventricle joined the incomplete and left-sided right ventricle through a muscular ventricular septal defect. The ventriculoarterial connections were concordant, with the aorta positioned posterior and to the right. The aortic valvar leaflets were in fibrous continuity with

Figure 3 Autopsy findings in case 3. (A) Isomerism of the morphologically right atrial appendages (RAAs), both of which were juxtaposed to the left of the arterial trunks. (B) Both atrial chambers emptied through a common atrioventricular valve (CAVV) into a ventricular mass showing right-hand topology, with dominance of the morphologically left ventricle (MLV). (C,D) The arterial trunks (AO, PT) arose in parallel fashion from the morphologically appropriate ventricles, with complete muscular infundibulums in both ventricles (dotted lines).

the leaflets of both atrioventricular valves, and the leaflets of the pulmonary valve were separated from the leaflet of the left atrioventricular valve by a muscular infundibulum. The arterial trunks exited from the heart in parallel fashion.

Case 6

This infant, failing to thrive, was admitted to hospital at age 7 months with cyanosis. Catheterisation showed a functionally single ventricle with double inlet, stenosis of the left

Figure 4 Autopsy findings in case 4. In this specimen, the right atrioventricular connection was absent (tricuspid atresia; findings not shown). (A) The left atrium communicated with the dominant left ventricle (MLV) through a mitral valve (MV), and there was fibrous continuity (arrowed) between the leaflets of the mitral and aortic (AO) valves. (B) The incomplete right-sided morphologically right ventricle (MRV), lacking its inlet component, gave rise to the pulmonary trunk (PT), being fed through a muscular ventricular septal defect (VSD).

atrioventricular valve and an atrial septal defect. Progressive congestive heart failure ensued, which did not respond to medical treatment. Catheterisation at age 21 months showed raised left atrial pressures. Atrial septectomy was carried out, along with banding of the pulmonary trunk. The child had severe cyanosis immediately after surgery, and was returned to the operating room for debanding, but did not survive.

Examination of this specimen showed a left-sided cardiac apex, the usual atrial arrangement, normally positioned atrial appendages, a defect within the floor of the oval fossa and double inlet to a dominant left ventricle through two separate atrioventricular valves. The incomplete right ventricle, positioned anteriorly and to the left, was connected by a muscular ventricular septal defect to the dominant ventricle. The ventriculoarterial connections were concordant, with parallel arterial trunks, but with the aorta posterior and to the right relative to the pulmonary trunk. The aortic valvar leaflets were in fibrous continuity with both atrioventricular valves, but the pulmonary valve was supported by a muscular infundibulum.

Case 7

This child presented with progressive cyanosis from birth. Catheterisation showed a right-sided heart with pulmonary atresia. She died at age 3 months, with severe desaturation and dyspnoea.

Examination of the heart showed a right-sided apex and isomerism of the morphologically left atrial appendages, with left juxtaposition of the right appendage. The venoarterial connections were normal, but the oval fossa was widely patent. The right atrioventricular connection was absent, with the left-sided atrium connected to a dominant morphologically left ventricle. The incomplete right ventricle, positioned anteriorly and to the right, was fed from the dominant ventricle through a muscular ventricular septal defect. The ventriculoarterial connections were concordant, with the arterial trunks arising in parallel fashion with the aorta left-sided and anterior relative to the pulmonary trunk. Both arterial valves were supported by complete muscular infundibulums. A major subpulmonary stenosis was seen, but flow to the aorta and aortic arch was unobstructed.

Case 8

This infant had cyanosis at birth and developed dyspnoea on exertion. Assessment at age 3 months suggested tricuspid atresia and pulmonary stenosis. She died of pneumonia at age 9 months.

Examination of the heart showed a right-sided apex, with the usual atrial arrangement but left juxtaposition of the right atrial appendage. Systemic and pulmonary venous connections were normal, but there was a large deficiency in the floor of the oval fossa. The right atrioventricular connection was absent, with the left atrium joined to a dominant morphologically left ventricle through a mitral valve. The rightward and anterior

morphologically right ventricle was incomplete, lacking its inlet component, and was fed through a muscular ventricular septal defect. The ventriculoarterial connections were concordant, with parallel arterial trunks, the aorta being left-sided and anterior relative to the pulmonary trunk. Both arterial valves were supported by an infundibulum, with subvalvar pulmonary stenosis and hypoplastic pulmonary arteries, but no obstruction to aortic flow. A single coronary artery arose from the right coronary aortic sinus. Some degree of myocardial non-compaction was noted in the dominant left ventricle.

Tables 1 and 2 summarise the salient anatomical findings in all eight hearts.

DISCUSSION

For many years, there was a doubt¹ regarding the several descriptions of arterial trunks arising from their appropriate ventricles, but with the aorta positioned anteriorly and to the left relative to the pulmonary trunk.^{9–11} At the time of these early descriptions, an anterior position of the aorta had been the criterion for describing the arterial trunks as being transposed; this arrangement, with the aorta positioned anteriorly, and hence transposed, yet still taking its origin from the morphologically left ventricle, was called anatomically corrected transposition. When it was finally established beyond doubt that the abnormal situation was a morphological reality, it was the inappropriate ventricular origins of the arterial trunks which was becoming accepted as the most appropriate definition for transposition; so the arrangement in which the arterial trunks arose from appropriate ventricles, but in abnormal relationships to each other, was called anatomically corrected malposition.¹ It was also argued, however, that hearts deemed suitable for this description should possess complete infundibulums in both ventricles. Other arrangements, in which the arterial trunks took origin from their appropriate ventricles, and exited from the base of the heart in a parallel fashion, as in the so-called anatomically corrected malposition, were denied this designation because the aortic valve, arising from the left ventricle, was in fibrous continuity with the leaflets of the mitral valve, or with one or both atrioventricular valves in the setting of double-inlet left ventricle. These arrangements were found when the morphologically right ventricle was itself left sided, or in the setting of mirror-imaged atrial arrangement but with the morphologically right ventricle in the right-sided position. As the pulmonary trunk was then to the left as in the normal heart, or to the right as expected for the mirror-imaged arrangement, it was argued that only the ventricular arrangement was abnormal. Hence, these unusual situations were described as isolated ventricular inversion,¹² isolated atrioventricular discordance or isolated ventricular non-inversion.¹³

Such argument, however, begs the question of the standard for determining the normal relationship of the arterial trunks. In the normal heart, the pulmonary trunk swings from right to

Table 1 Characteristics of the ventricles and ventriculoarterial junctions

Case	Position of RV	Infundibulum	Position of aorta	Outflow obstruction	Coronary arteries
1	L	Subpulmonary	R and P	Hypoplastic AoV and ascending aorta	Single RCA
2	R	Bilateral	L and A	No obstruction	Normal
3	R	Bilateral	L and A	Pulmonary subvalvar and valvar	Single RCA
4	R and A	Subpulmonary	L and A	Pulmonary subvalvar and valvar	Single RCA
5	L and A	Subpulmonary	R and P	No obstruction	RCA, LCA from RCC
6	L and A	Absent	R and P	No obstruction	Normal
7	R and A	Bilateral	L and A	Pulmonary subvalvar and valvar	Normal
8	R and A	Bilateral	L and A	Pulmonary subvalvar and valvar	Single RCA

A, anterior; AoV, aortic valve; L, left; LCA, left coronary artery; P, posterior; R, right; RCA, right coronary artery; RCC, right coronary cusp; RV, right ventricle.

Table 2 Segmental analysis and associated features

Case	Apex	Atriums	Venous return	IAS	Atrioventricular connection	RV	LV
1	L	UAA	Normal	Intact	Biv, discordant	Left hand	
2	L	UAA	Normal	Intact	Biv, concordant, hypoplastic MV	Right hand	Hypoplastic
3	R	RIAA, UAA	Bilateral SCV, ICV to RA TAPVD to LSCV	Secundum ASD	Biv, ambiguous, common AVV	Right hand Hypoplastic	
4	R	UAA, UAA	LSCV to CS	Secundum ASD	Univ, absent right	Incomplete	Dominant
5	L	UAA	TAPVD to CS	Secundum ASD	Univ, DILV	Incomplete	Dominant
6	L	UAA	Normal	Secundum ASD	Univ, DILV	Incomplete	Dominant
7	R	UAA, UAA	?	Secundum ASD	Univ, absent right	Incomplete	Dominant
8	R	UAA, UAA	?	Common atrium	Univ, absent right	Incomplete	Dominant, LVNC

ASD, atrial septal defect; Biv, biventricular; CS, coronary sinus; DILV, double-inlet left ventricular; IAS, intra-arterial septum; ICV, inferior caval vein; L, left; UAA, left isomerism of the atrial appendages; UAA, left juxtaposition of the right atrial appendage; LSCV, left superior caval vein; LV, left ventricle; LVNC, left ventricular non-compaction; MS, muscular; MV, mitral valve; PM, perimembranous; R, right; RA, right atrium; RIAA, right isomerism of the atrial appendages; RV, right ventricle; SCV, superior caval vein; TAPVD, total anomalous pulmonary venous drainage; UAA, usual atrial arrangement; Univ, univentricular; VSD, ventricular septal defect; ?, the venous connections could not be assessed.

left as it exits from the morphologically right ventricle, and then spirals to the left round the ascending part of the aorta as it branches into the right and left pulmonary arteries. In the setting of the mirror-imaged normal heart, in contrast, the mirror-imaged but otherwise normally arranged pulmonary trunk swings from left to right as it exits the base of the ventricular mass, and then spirals to the right as it turns round the aorta to reach its bifurcation. It could be argued, therefore, that when there is left-handed ventricular topology, the criterion for normal relationships of the aorta and pulmonary trunk should be the situation as seen in the mirror-imaged but normal heart. Similarly, when seen in the setting of right-handed ventricular topology, then the typical pattern of normal relationships should be considered to be the normal default option. When considered in this light, the relationships of the arterial trunks, neither as seen in the isolated ventricular inversion nor as seen in the isolated ventricular non-inversion, can be considered to be normal. This is the more so, as the arterial trunks themselves exit in parallel rather than spiral fashion as they leave the base of the ventricular mass. A case can be made, therefore, for including all of these abnormal arrangements at the ventriculoarterial junctions in the same group as the so-called anatomically corrected malposition.

This case is strengthened when we note perhaps the most important anatomical principle to emerge during the 20th century. This was the emphasis given by Van Praagh *et al*¹⁴ to the “morphological method”, which rightly stated that one variable morphological feature should not be determined on the basis of another feature which is itself variable. When considering the description of arterial trunks exiting from the heart in parallel fashion, application of this important principle dictates that similar arrangements should not be described in different fashion simply because, in some instances, there is fibrous continuity between the leaflets of an atrioventricular and an arterial valve. Therefore, it seems justifiable to group together all the hearts described in our review. This is because (1) the arterial trunks arise from their morphologically appropriate ventricles and (2) they exit from the base of the ventricular mass in a parallel rather than spiralling fashion. The variability found in infundibular morphology is but one of the morphological features that differ among the group of examined hearts. It is equally noteworthy that, among our hearts, we found all possible types of variability at the atrioventricular junctions—namely, concordant, discordant, ambiguous and biventricular—and double-inlet connections, along with hearts showing the absence of one atrioventricular connection. We also found hearts with the usual arrangement and both types of isomeric atrial appendages, as well as hearts with two atrioventricular valves as opposed to a common or a solitary atrioventricular valve. Also, finding the coronary

arteries arising exclusively from the right coronary aortic sinus, either through a solitary artery or two separate arterial orifices, in five of eight specimens must surely be more than coincidence, considering the rarity of these segmental combinations.

The rarity of these lesions reflects a lack of appreciation of their nature, or else lack of attention thus far to the parallel as opposed to the spiralling arrangement of the arterial trunks as they exit from the ventricular mass. Although we discovered our eight described examples in the archive of congenitally malformed hearts in Toronto, when we also searched our extensive echocardiography database, established in the mid-1980s, we found no examples of these particular arterial arrangements. This could simply be because no such patients presented for assessment or, perhaps, because those making the diagnosis failed to code the abnormal arterial relationships, and infundibular morphologies of patients seen during this period. The association between the so-called anatomically corrected malposition and tricuspid atresia is well established¹⁵; yet, no patients with tricuspid atresia in our database had been coded as having this arrangement. Only when attention is given to the ventricular origin of the arterial trunks and also to the infundibular morphologies and the relationships of the intrapericardial arterial trunks can we establish the true prevalence of these unusual and fascinating arrangements at the ventriculoarterial junctions.

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IMAGES IN CARDIOLOGY

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Massive intrapericardial fibroma presenting with palpitations

A 52-year-old woman, with no previous past medical history of note, presented with a six-month history of palpitations and raised jugular venous pressure (JVP). A chest radiograph revealed a large mass adjacent to the right atrium (panel A) which was subsequently confirmed on transthoracic echocardiography. A computed tomographic (CT) scan demonstrated a 15 cm heterogeneous mass of indeterminate origin and extent. Further imaging using magnetic resonance revealed the mass to be arising from the atria (most likely the right atrium) (panel B) and the appearances were consistent with those of a giant myxoma. Furthermore, while there was no evidence of local invasion, compression of the left upper pulmonary vein was evident. Operative exploration was carried out through a median sternotomy. The mass was intrapericardial, lying in the right posterolateral pericardial space, compressing and deforming the right and left atrium. Intraoperative transoesophageal echocardiography confirmed the constrictive effect of this extra-cardiac mass upon the right and left atria. A preliminary biopsy revealed a benign fibromatous tumour. This was excised one week later through a right thoracotomy without the need for cardiopulmonary bypass (panel C). Upon removal of the mass, haemodynamic parameters returned to normal. The patient made an uneventful postoperative recovery and was discharged home on the sixth postoperative day. Histological examination of the mass confirmed a benign fibroma that was completely excised. Staging using clinical and radiological means was invaluable in ensuring successful resection of this rare tumour.

(A) Chest radiograph demonstrating a large mass silhouetting with right heart border. (B) Magnetic resonance image (MRI) revealing a mass arising from the atria. (C) Visualisation of fibroma following right thoracotomy and incised surface of fibroma. (D) Postoperative MRI following the removal of extrinsic compression upon the left and right atria.

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