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DOI:

[10.1053/j.semtcvs.2020.05.016](https://doi.org/10.1053/j.semtcvs.2020.05.016)

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*Document Version*

Peer reviewed version

*Citation for published version (Harvard):*

Maria, N, Roderick, M, Stickley, J, Drury, N, Jones, T, Barron, D, Brawn, W, Anderson, RH & Crucean, A 2020, 'Anatomical studies of transposition – an argument for a unifying morphological classification', *Seminars in Thoracic and Cardiovascular Surgery*, vol. 32, no. 4, pp. 920-926. <https://doi.org/10.1053/j.semtcvs.2020.05.016>

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**Anatomical studies of transposition – an argument for a unifying morphological classification**

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Key words: Discordant ventriculo-arterial connections; Taussig-Bing malformation; Nomenclature; Atrial arrangement; d-transposition

Abstract word count: 236

Manuscript word count: 3500

Number of references: 14

Number of tables: 1

Number of figures and legends: 6

**Conflict of interest statement:** There is no conflict of interest for any of the authors.

**Source of funding:** No specific funding was required for designing and producing this study. Nigel Drury is funded by an Intermediate Clinical Research Fellowship from the British Heart Foundation [FS/15/49/31612] and the Institute of Cardiovascular Sciences is supported by an Accelerator Award from the British Heart Foundation [AA/18/2/34218].

36 **Glossary**

37 DORV - Double outlet right ventricle

38 IVS - Intact ventricular septum

39 TGA - Transposition of the great arteries

40 VSD - Ventricular septal defect

41

42

43 RHA – Robert H. Anderson

44 AC – Adrian Crucean

45 NM – Nikita Maria

46 MER – Max E. Roderick

47 NED – Nigel E. Drury

48 TJJ – Timothy J. Jones

49 DJB – David J. Barron

50 WJB – William J. Brawn

51

52 **Central Message**

53        Descriptions of patients with transposition must go beyond “d-transposition” to include  
54    important anatomical information, such as arterial trunk arrangement, which may influence  
55    surgical management.

56 **Perspective statement**

57       Patients with concordant atrioventricular and discordant ventriculo-arterial connections  
58 have classically been described as having “d-transposition”, but this may not be accurate.  
59 Further details regarding arterial trunk relationships, origins of coronary arteries, infundibular  
60 morphology and phenotypic variations of ventricular septal defects are needed to guide optimal  
61 surgical management.

**Abstract:**

**Objectives:** In the setting of transposition, recognition of the variability in the morphology of the outflow tracts and the arterial trunks is essential for surgical repair. Presence of a unifying system for classification would minimize the risk of misunderstanding.

**Methods:** We examined an archive of 142 unrepaired hearts with transposition, defined as the combination of concordant atrioventricular and discordant ventriculo-arterial connections, combined with the clinical records of 727 patients undergoing the arterial switch procedures.

**Results:** In the setting of usual atrial arrangement, we found the aortic root in leftward or posterior location in up to 5% of our cohorts, making the common term ‘d-transposition’ ambiguous. Variability in the relationship of the trunks was commoner in the setting of deficient ventricular septation, especially when the trunks were side-by-side (14% vs 5.5% when the ventricular septum was intact). Bilateral infundibulums were present in 14% of cases, and bilaterally deficient infundibulums in 3.5%. Both of these findings were more common with deficient ventricular septation. Fibrous continuity between the tricuspid and pulmonary valves was not always seen with perimembranous defects, particularly when there were bilateral infundibulums. Fibrous continuity between the leaflets of the tricuspid and mitral valves, in contrast, proved a unifying characteristic of the perimembranous defect.

**Conclusion:** The combination of concordant atrioventricular and discordant ventriculo-arterial connections is best described simply as ‘transposition’, with precision provided when details are given of atrial arrangement and the relationships of the arterial trunks.

Word count: 236

## **Introduction**

Transposition, when used in isolation, describes the combination of concordant atrioventricular and discordant ventriculo-arterial connections<sup>1</sup>. This segmental combination can occur with a variety of other lesions, the most common of which is a ventricular septal defect. There can also be considerable variability in the exact relationship of the so-called great vessels, which are described more specifically as the arterial trunks. The pattern of the coronary arteries also shows significant variability<sup>2</sup>. Despite detailed previous analyses, the variability in the morphology of the outflow tracts and the relationship of the arterial trunks has not always been well described. An understanding of these features, including the specific phenotypic features of a ventricular septal defect, if present, is essential for the surgeon in planning accurate surgical repair. It would be optimal, therefore, if a common language could be used for describing the variations. In this regard, when defined as the combination of concordant atrioventricular and discordant ventriculo-arterial connections, transposition is almost always found with usual atrial arrangement. On very rare occasions, nonetheless, it can be encountered in the mirror-imaged format<sup>3</sup>. When found in the latter setting, there is left-handed ventricular topology, and the transposed aorta is typically left-sided<sup>3</sup>. Even when found with usual atrial arrangement, however, the aorta can be left-sided, or even posterior. The finding of these alternative segmental combinations calls into question the common practice of describing the entity as “d-transposition”.

Irrespective of such semantic controversies, the prognosis for patients born with transposition has been transformed over the last half-century. Initially repaired surgically by redirecting the atrial venous returns, using either the Mustard or the Senning procedures, it is now customary to achieve surgical repair by the arterial switch operation. There are, nonetheless, multiple potential anatomical variations. These include variability in the relationships of the intrapericardial arterial trunks, differences in infundibular morphology, and

the presence of associated defects, such as ventricular septal defects or obstruction of the ventricular outflow tracts. The outstanding results of the arterial switch in the modern era are such that attention is increasingly focused on improving outcomes for the more unusual variants in morphology, such as unusual relationship of the arterial trunks, or intramural coronary arterial patterns. Attention is also focused on optimizing surgical repair to minimize morbidity and late-sequels, such as post-operative heart block or late obstruction of the outflow tracts. These problems are best addressed on the basis of complete understanding of the morphology. In the Birmingham Archive of congenitally malformed hearts, there are 142 hearts with transposition. Assessment of these specimens provided an opportunity to identify particular anatomic findings that might impact on future surgical morbidity. These findings provided a focus for similar assessment of our clinical database.

## **Methods:**

The archive of autopsied hearts at Birmingham Children's Hospital contains around 2000 specimens, collected since 1939. The majority were entered into the archive in the periods when there was little, if any, surgical treatment for transposition. Of the specimens in the archive, 138 have the combination of concordant atrioventricular and discordant ventriculo-arterial connections. An additional 4 hearts show comparable features, but with the pulmonary trunk overriding an interventricular communication in the setting of double outlet right ventricle. These are examples of the so-called Taussig-Bing arrangement, when this lesion is defined as suggested by Lev and colleagues on the basis of overriding of the ventricular septum by the pulmonary trunk, with either discordant ventriculo-arterial connections or double outlet right ventricle according to the precise commitment of the overriding trunk.<sup>4</sup>. All these hearts have usually arranged atrial appendages, or "situs solitus". Our clinical database consists of 727 patients who have undergone the arterial switch procedure at Birmingham Children's Hospital between 1988 and 2017. By combining these cohorts, the focus of our study was,



134 firstly, to define the frequencies and variation in the arrangements of the arterial trunks and  
135 infundibular morphology. Our second aim was to focus on the more unusual morphological  
136 variants, hoping to establish a reliable and consistent method of classification which would be  
137 applicable to all.

138         Having identified the hearts fulfilling the diagnosis of transposition during the initial  
139 archiving of the catalogue (RHA, AC), along with the hearts catalogued as showing the  
140 Taussig-Bing arrangement with the ventriculo-arterial connection of double outlet right  
141 ventricle, we obtained post-mortem reports for each heart, providing the necessary clinical  
142 background. Each heart was then analyzed by a primary and secondary observer (NM, MER),  
143 compared to the post-mortem account, and reviewed again by AC. We assessed the segmental  
144 combinations, the arterial relationships, the infundibular morphologies, and all associated  
145 malformations. Assessment of the patterns of the coronary arteries will form the basis of a  
146 separate submission. Data were analyzed using Microsoft Excel, using Fisher's exact test of  
147 independence to establish statistical significance, since the data sets were deemed too small to  
148 use the Chi-squared test. Statistical significance was presumed when the p value was less than  
149 0.05. In collaboration with our surgical colleagues (NED, TJJ, DJB, WJB), we accessed the  
150 details of the patients entered into the clinical database. In this regard, we based our description  
151 of the anatomical findings on the notes made by the operating surgeon subsequent to the switch  
152 procedure.

## 153 **Results:**

154         Of the 142 hearts catalogued within the archive as having transposition or the Taussig-  
155 Bing malformation, all with usual atrial arrangement, the ventricular septum was intact in 67.  
156 In the remaining 75 hearts, which included 5 having the discordant end of the Taussig-Bing  
157 malformation as defined by Lev and associates<sup>4</sup>, and the 4 hearts with the double outlet right

ventricle variant,<sup>4</sup> there was at least one channel between the ventricles. The channel is better described as an interventricular communication in the setting of double outlet, since it is not closed by the surgeon during the operative repair, but rather tunneled to the pulmonary root to commit the root to the left ventricle as the neo-aortic outflow tract subsequent to the arterial switch procedure<sup>5</sup>. We had excluded any hearts with significant hypoplasia or atresia of the left outflow tract, as these would not have been suitable for the arterial switch operation, and hence outside the focus of our study.

#### Relationship of the Arterial Trunks:

In 122 of the 138 hearts (88%), the aorta was positioned anteriorly relative to the pulmonary trunk. The trunks were side-by-side in 13 (9.5%), with the aortic trunk found in posterior position in 3 (2.5%). When the aorta was anterior, it was positioned to the right in 93 hearts (67%). The aorta was deemed to be positioned in directly anterior position relative to the pulmonary trunk in 25 hearts (18%), while in an additional 4 hearts (3%), the aorta was anterior and leftward, 3 of these found in the presence of a ventricular septal defect. When the hearts were further analyzed according to integrity of the ventricular septum, significant differences emerged in the frequency of the patterns of relationship of the arterial trunks (Figure 1). An anteriorly located aorta was more frequent when the ventricular septum was intact, occurring in 75% of cases, compared to 57% in the hearts with deficient septation ( $p=0.01$ ). The arterial trunks were more frequently side-by-side in the hearts with ventricular septal defects as opposed to those with an intact ventricular septum, at 14% vs 5.5% (see Figure 1). Posterior and rightward aortas were found in three hearts, 2 associated with a ventricular septal defect, with all having right-handed ventricular topology. This is so-called “normal relations”, but with discordant ventriculo-arterial connections, since in all hearts the pulmonary trunk arose exclusively from the left ventricle.

### Infundibular Morphology:

The observed variability in infundibular morphology is summarized in Table 1. The most common finding was presence of a sub-aortic infundibulum in the right ventricle, with fibrous continuity in the roof of the left ventricle between the leaflets of the pulmonary and mitral valves, seen in 132 of the hearts (93%). Bilateral infundibulums were found in 20 hearts (14.1%), with 14 of these having deficient ventricular septation, including three of the four hearts with the ventriculo-arterial connection of double outlet right ventricle. Bilaterally deficient infundibulums were found in 5 hearts (3.5%). Juxta-arterial and doubly committed ventricular septal defects were found in three of these hearts (Figure 2). In one of the hearts with double outlet right ventricle, there was also bilateral deficiency of the infundibulums, in this instance with fibrous continuity between the leaflets of the aortic and tricuspid valves, and also between those of the mitral and pulmonary valves. In the final heart, which has a posterior aorta, there was a sub-pulmonary infundibulum, but fibrous continuity in the roof of a ventricular septal defect between the leaflets of the aortic and mitral valves.

### Morphology of the Ventricular Septal Defect:

Surgical repair had been attempted in 11 of the hearts, making assessment of the defect impossible because of the materials and sutures used during closure. We excluded these hearts from subsequent analysis. In the remaining 60 hearts, the defect was perimembranous in 40 (67%), muscular in 17 (28%), and doubly-committed and juxta-arterial in the remaining 3 (5%). Multiple defects were found in five hearts. These were all muscular in three, with combined perimembranous and muscular defects found in two. In the hearts with perimembranous and muscular defects, the conduction axis ran through the muscle bar separating the perimembranous defect from the muscular defect, which opened to the inlet of the right ventricle. In all the perimembranous defects, there was fibrous continuity postero-

inferiorly between the leaflets of the mitral and tricuspid valves. In the majority of these hearts, there was also fibrous continuity between the leaflets of the tricuspid and pulmonary valves in the roof of the defect (Figure 3). In a minority, nonetheless, the muscular outlet septum formed the cranial margin of the defect, producing discontinuity between the leaflets of the tricuspid and pulmonary valves (Figure 4). Despite the muscular interposition between the pulmonary and tricuspid valves, the conduction axis remained postero-inferior relative to the margins of the defect, an important consideration for those undertaking surgical repair.

In the hearts with perimembranous defects, the defect opened primarily to the inlet of the right ventricle in 17.5%, to the outlet in 50%, and opened centrally in the remaining 32.5%. In two of the hearts with the defect opening to the inlet, there was malalignment between the atrial septum and the muscular ventricular septum, with straddling and overriding of the tricuspid valve (Figure 5). In this setting, the atrioventricular conduction axis remains inferior, but is unable to arise from the regular atrioventricular node positioned at the apex of the triangle of Koch. Instead, the axis is carried on the malaligned ventricular septum, arising from an anomalous inferior atrioventricular node<sup>6</sup>.

#### Comparison with Clinical Database:

Of the patients making up the clinical database, 424 (58.3%) had an intact ventricular septum, with 303 patients (41.7%) having deficient ventricular septation. Included in the latter group were 25 patients diagnosed, on the basis of the definition as provided by Lev and colleagues<sup>4</sup>, as having the Taussig-Bing variant of double outlet right ventricle. The proportion of patients with deficient ventricular septation was not statistically significantly different ( $p = 0.22$ ) from the comparable findings from the cardiac archive. The incidence of double outlet was also similar between the clinical database and the archive. In the clinical database, however, a greater proportion of the patients was diagnosed as having defects with exclusively

muscular margins, this feature being found in 134 patients (44.2%), contrasting with the archive, where 59.7% of defects were perimembranous. Perimembranous defects were found in 138 patients (45.5%), with 11 of these patients having co-existent muscular defects. Doubly committed defects were diagnosed in 7 patients (2.3%). The incidence of all the phenotypic variants, therefore, was similar to those found in the archive. However, we accept that it is sometimes difficult to precisely define such anatomic details in the midst of a challenging operation. The type of defect had not been coded in 9% of the patients. Straddling and overriding of the tricuspid valve was recognized in three patients, with straddling of the mitral valve in one patient. In terms of clinical outcomes, it was noted that 5 patients (0.7%) developed post-operative heart-block requiring pacemaker implantation; two had transposition with deficient ventricular septation (one perimembranous outlet defect, one muscular inlet defect) and three had Taussig-Bing malformation with a subpulmonary defect, one of whom had a straddling mitral valve. The only major difference found between the clinical database and the archive was the finding of 6 patients (0.8%) with mirror-imaged atrial arrangement, all also exhibiting left-handed ventricular topology. In this small group of patients, the aorta was described as being directly anterior to the pulmonary trunk in 3, anterior and to the right in 2, and anterior and to the left in only one.

## **Discussion:**

The morphological characteristics of transposition are complex, and a common language to classify and describe these morphological variants is fundamental. It clarifies the understanding of the condition and its variants such that surgical repair can be tailored to the individual anatomy. In this regard, a morphologic archive provides a unique resource to study all the potential variations. A clinical database then provides the opportunity to assess the clinical relevance of the different morphological patterns on the surgical outcomes, serving to highlight the areas of greater risk and complexity. Some morphological details, nonetheless,

are not always coded in clinical databases, leaving gaps in our knowledge. We were able to use both a large archive and a clinical database from the same hospital. This permitted us to combine lessons learned from both sources, comparing one against the other, and focusing on morphological relationships that may not have been appreciated previously. Features such as infundibular morphology, for example, have not always been recorded by surgeons performing the arterial switch. The improvements in pre-operative imaging should now make this feature easier to document. The feature may well prove to be significant in determining the incidence of dilation of the new aortic root after the arterial switch procedure, as well as influencing potential correction using the Nikaidoh procedure.

Our study has focused on the variations in infundibular morphology, the relationships of the arterial trunks, and when present, the anatomic features of channels between the ventricles. Figure 6 illustrates the arterial trunk relationships found overall within the archive. The findings confirm that the anterior and rightward aorta is much more common in the setting of an intact ventricular septum. Side-by-side arterial trunks, in contrast, are much more common when the ventricular septum is deficient. A sub-aortic infundibulum in the right ventricle, with continuity between the leaflets of the pulmonary and mitral valves in the roof of the left ventricle, was the most common infundibular morphology, found in over four-fifths of the cohort, and more common when the ventricular septum was intact. Bilateral infundibulums, found in one-fifth of cases, were more common in association with deficient ventricular septation, including patients with the double outlet variants of the Taussig-Bing lesion. Half of those with bilateral infundibulums, nonetheless, had discordant ventriculo-arterial connections, accounting for nearly one-tenth of the hearts obtained from patients with an intact ventricular septum. It will be of interest to follow-up such patients over the long term to establish whether this feature does, indeed, have any influence on the integrity of the neo-aortic valve and the function of the new aortic root.

One of our most important findings was the recognition that fibrous continuity between the leaflets of the tricuspid and pulmonary valves did not serve a diagnostic feature for perimembranous defects, particularly in the setting of bilateral infundibulums. The consistent finding was fibrous continuity between the leaflets of the mitral and tricuspid valves in the postero-inferior border of the defects. This was always present, regardless of infundibular morphology. This feature, therefore, can guide the surgeon to the anticipated position of the conduction axis when closing such defects, thus minimizing the risk of inducing heart block.

Bilateral deficiency of the infundibulums was unusual, but occurred with similar frequency irrespective of the integrity of the ventricular septum. This association has not usually been described with an intact ventricular septum, although it did not appear to have any specific clinical implications. When the ventricular septum was deficient, the deficient infundibulums were usually found with doubly-committed and juxta-arterial defects, which are usually more easily approached surgically via the aortic valve<sup>7</sup>. The defects were perimembranous in almost two thirds of the archived hearts, in keeping with the findings from previous studies<sup>8,9</sup>.

The noted variabilities in relationships of the arterial trunks call into question the continuing tendency to describe the overall combination of segmental connections as “d-transposition”. All of the hearts found in the archive had usual atrial arrangement and right-handed ventricular topology. Even in these hearts, a significant number had the aorta positioned directly anterior to the pulmonary trunk, while in a small proportion, the aortic root was located leftward. We also found hearts exhibiting so-called “posterior” transposition<sup>10,11</sup>. These hearts cannot readily be categorized using the codes currently available in the system now accepted for the eleventh iteration of the International Classification of Disease<sup>12</sup>. In Van Praaghian notation, the hearts with usual atrial arrangement and leftward aorta would be described as showing transposition {S,D,L}<sup>13</sup>. Indeed, in the “segmental approach” as introduced by Van

Praagh,<sup>13</sup> the information regarding the relationship of the arterial roots was included as part of the segmental notation, rather than as a prefix attached to the specific lesion. The findings call into question, therefore, the common practice of describing all patients with the most common variant of discordant ventriculo-arterial connections as exhibiting “d-transposition”. In the clinical database, 6 of the patients undergoing the arterial switch operation had mirror-imaged atrial arrangement and left-handed ventricular topology. In Van Praaghian terms, these patients are expected to have transposition {I,L,L}<sup>14</sup>. Surprisingly, the surgeons reported the aorta to be left-sided in only one of the six. In patients with mirror-imagery, therefore, some can have right-sided aortas, albeit not with right-handed ventricular topology. Taken overall, our findings surely indicate that the time has come to cast aside the penchant for describing patients with concordant atrioventricular and discordant ventriculo-arterial connections as having “d-transposition”. The patients can simply be described as having transposition. To provide the details needed for optimal individual treatment, it is necessary to account for the atrial arrangement, ventricular topology, the relationship of the arterial trunks, the origin of the coronary arteries, the details of infundibular morphology, and the phenotypic variations of ventricular septal defects. This approach then serves to provide unambiguous and complete information.



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## Tables

Table 1. The incidence of infundibular morphology across the hearts. Key: \*including double outlet right ventricle variants; \*\*loss of pulmonary-mitral continuity. Abbreviations: IVS - intact ventricular septum; VSD – ventricular septal defect; TGA – transposition of great arteries.

Morphology	TGA/IVS	TGA/VSD*	Total
Single, Sub-Aortic Infundibulum	88.0%	73.0%	81.0%
Single, Sub-Pulmonary Infundibulum	1.3%	1.5%	1.4%
Double Infundibulums**	8.0%	20.9%	14.1%
Absent Infundibulum	2.7%	4.5%	3.7%

## Legends

Graphical abstract legend: A study looking at incidence of anatomical morphologies in hearts with transposition.

Central Picture legend: Incidence of various arterial trunk relationships observed within the archive.

Figure 1. Relationship of the great vessels seen in patients with transposition and intact septum and those with associated ventricular septal defects. (TGA – transposition of the great arteries, IVS – intact ventricular septum, VSD – ventricular septal defect, Ao – aorta, PA – pulmonary trunk).

Fig 2. The image shows the right ventricular aspect of a juxta-arterial ventricular septal defect. It opens to the right ventricle between the limbs of the septomarginal trabeculation (yellow Y). A muscular postero-inferior rim will protect the atrioventricular conduction axis should surgical correction be attempted in such hearts. The roof of the defect is formed by fibrous continuity between the leaflets of the aortic and pulmonary valves.

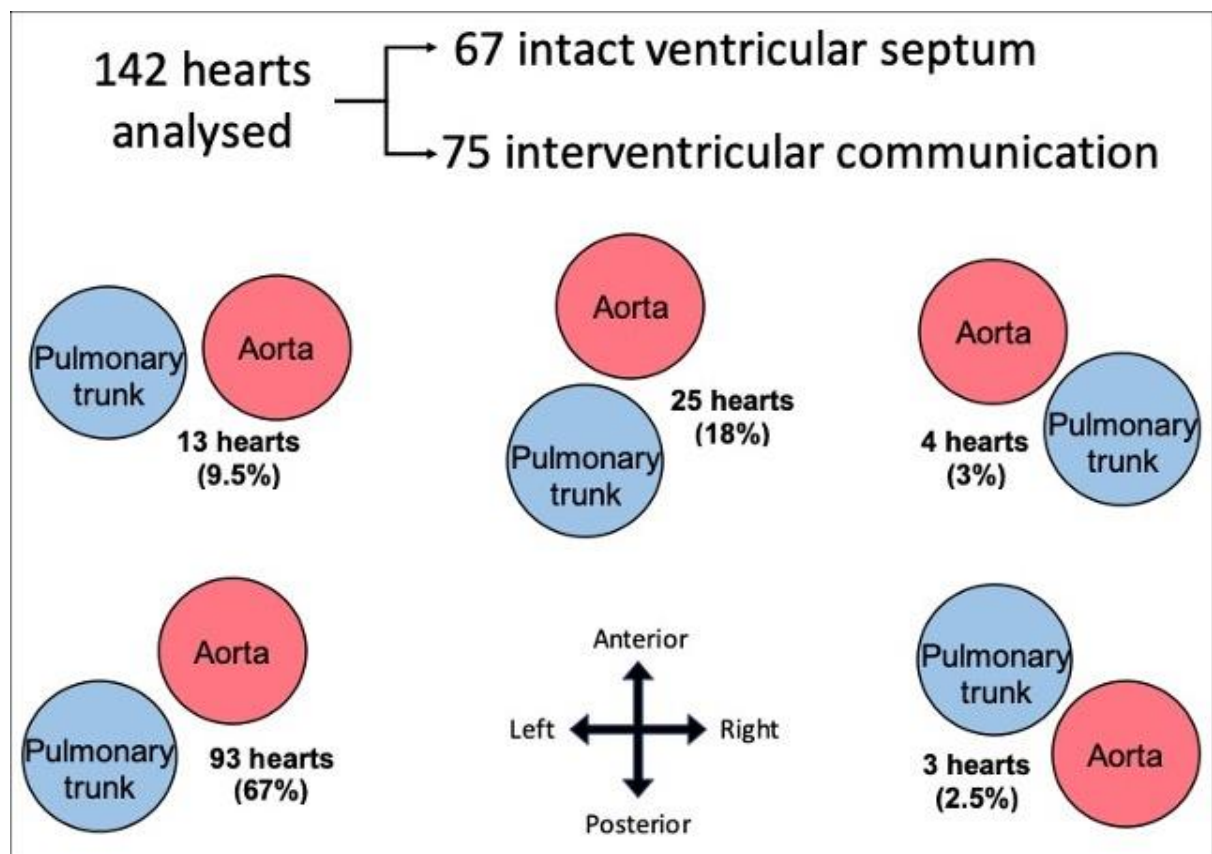
Figure 3. The image shows the phenotypic features of a perimembranous ventricular septal defect opening to the inlet of the right ventricle. There is fibrous continuity between the leaflets of the mitral and tricuspid valves, and also between the leaflets of the tricuspid and pulmonary valves. The atrioventricular conduction axis will be at risk postero-inferiorly in this setting.

Figure 4. The images show a heart with a perimembranous ventricular septal defect (VSD) viewed (A) from the right ventricle and (B) from the left ventricle. In this heart, as seen in panel A, the defect opens to the outlet of the right ventricle between the limbs of the septomarginal trabeculation (yellow Y). Fibrous continuity, however, exists only between the leaflets of the mitral and tricuspid valves. As seen in panel B, viewed from the left ventricle, the muscular outlet septum interposes between the leaflets of the pulmonary and tricuspid valves. There is

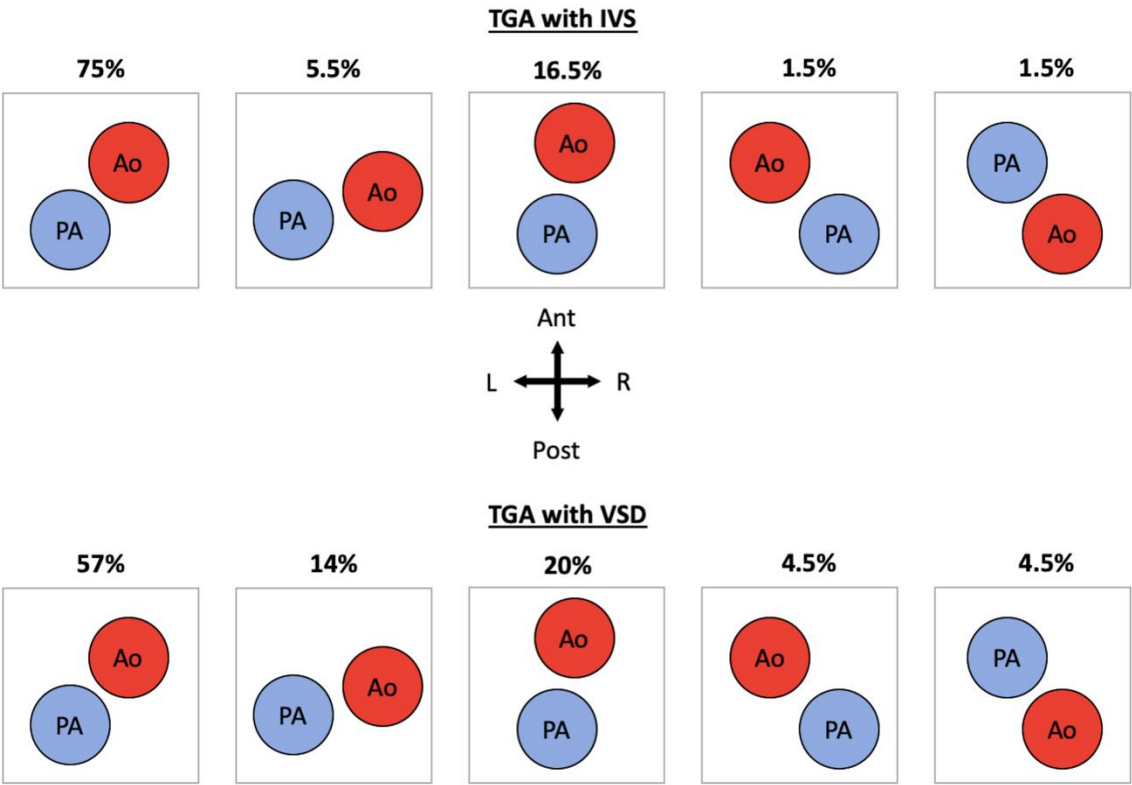
still fibrous continuity, nonetheless, between the leaflets of the mitral and tricuspid valves, which incorporates the transilluminated central fibrous body. Because of this feature, the conduction axis remains at risk postero-inferiorly.

Figure 5. The image shows the right ventricular aspect of a perimembranous defect opening to the inlet of the right ventricle, but in the setting of straddling and overriding of the tricuspid valve. The conduction axis in this setting (red bar) remains postero-inferior, but no longer arises from the regular atrioventricular node at the apex of the triangle of Koch (white star with red borders). Instead, the axis, carried on the malaligned ventricular septum, arises from an anomalous inferior atrioventricular node (red circle with white borders).

Figure 6. Incidence of various arterial trunk relationships observed within the archive, highlighting that, overall, an anterior and rightward aorta was the most commonly observed position.



Central Picture legend: Incidence of various arterial trunk relationships observed within the archive.

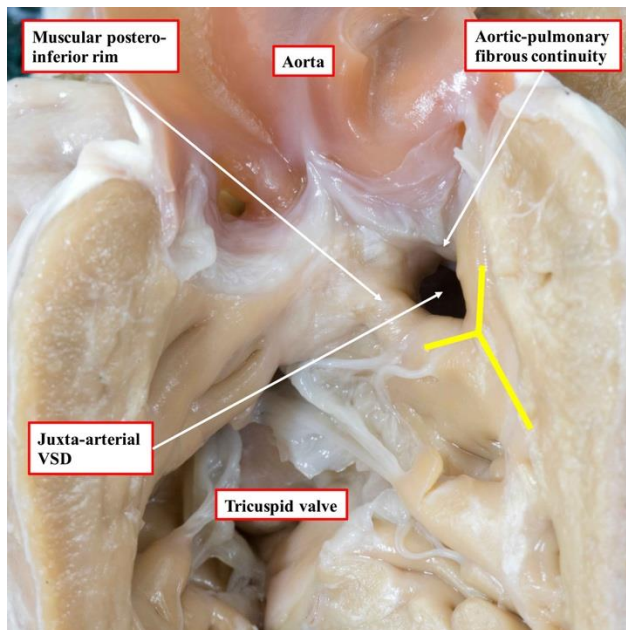


414

415 Figure 1. Relationship of the great vessels seen in patients with transposition and intact septum  
416 and those with associated ventricular septal defects. (TGA – transposition of the great arteries,  
417 IVS – intact ventricular septum, VSD – ventricular septal defect, Ao – aorta, PA – pulmonary  
418 trunk).

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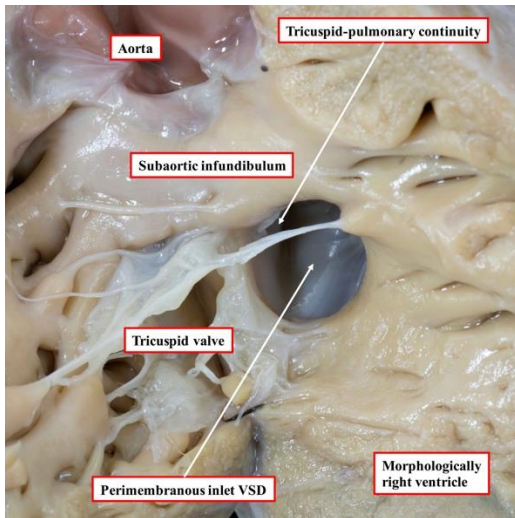
421

422 Figure 2. The image shows the right ventricular aspect of a juxta-arterial ventricular septal  
423 defect. It opens to the right ventricle between the limbs of the septomarginal trabeculation  
424 (yellow Y). A muscular postero-inferior rim will protect the atrioventricular conduction axis  
425 should surgical correction be attempted in such hearts. The roof of the defect is formed by  
426 fibrous continuity between the leaflets of the aortic and pulmonary valves.

427



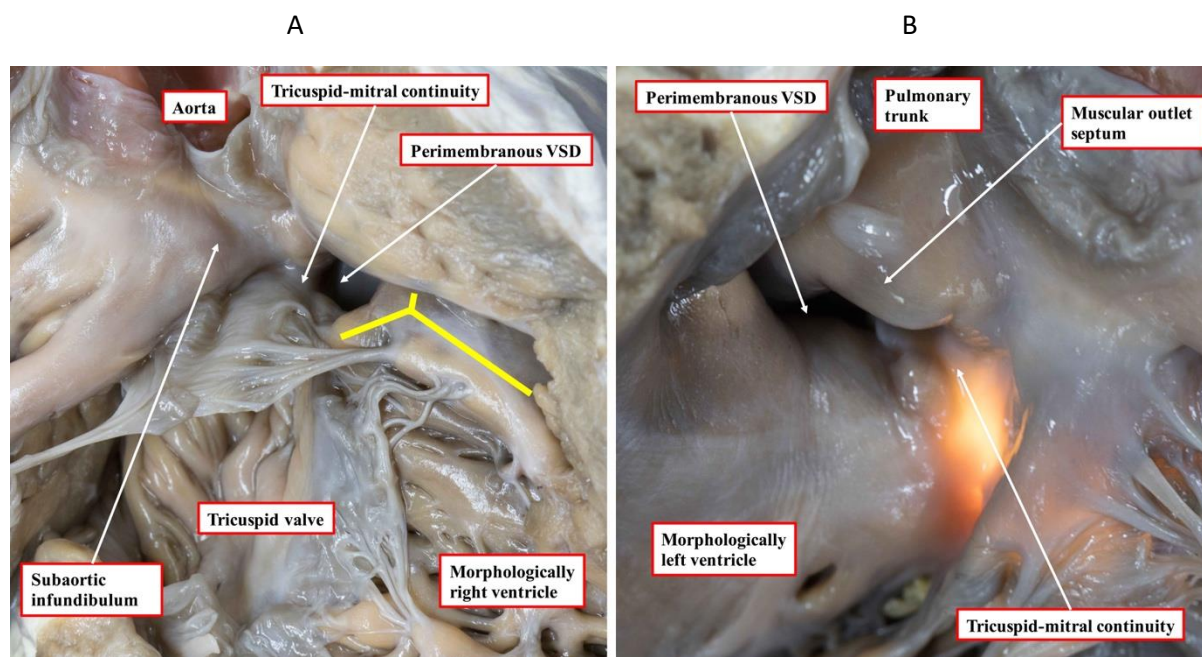
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429

430 Figure 3. The image shows the phenotypic features of a perimembranous ventricular septal  
431 defect opening to the inlet of the right ventricle. There is fibrous continuity between the leaflets  
432 of the mitral and tricuspid valves, and also between the leaflets of the tricuspid and pulmonary  
433 valves. The atrioventricular conduction axis will be at risk postero-inferiorly in this setting.

434

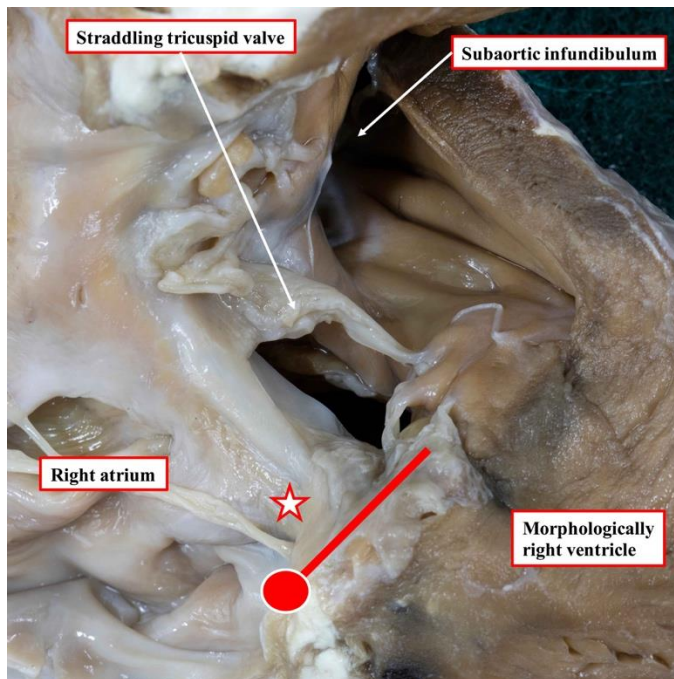


436

437 Figure 4. The images show a heart with a perimembranous ventricular septal defect (VSD)  
 438 viewed (A) from the right ventricle and (B) from the left ventricle. In this heart, as seen in panel  
 439 A, the defect opens to the outlet of the right ventricle between the limbs of the septomarginal  
 440 trabeculation (yellow Y). Fibrous continuity, however, exists only between the leaflets of the  
 441 mitral and tricuspid valves. As seen in panel B, viewed from the left ventricle, the muscular  
 442 outlet septum interposes between the leaflets of the pulmonary and tricuspid valves. There is  
 443 still fibrous continuity, nonetheless, between the leaflets of the mitral and tricuspid valves,  
 444 which incorporates the transilluminated central fibrous body. Because of this feature, the  
 445 conduction axis remains at risk postero-inferiorly.

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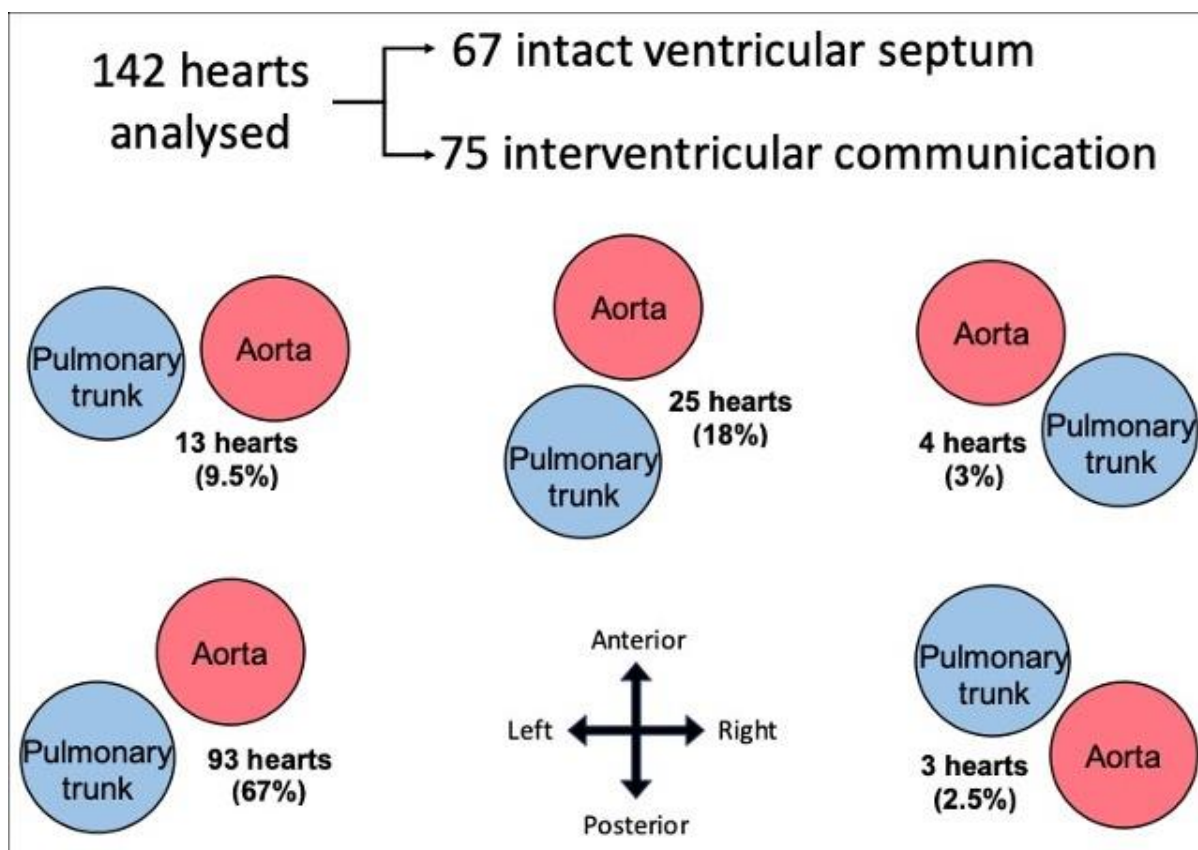


448

449 Figure 5. The image shows the right ventricular aspect of a perimembranous defect opening to  
450 the inlet of the right ventricle, but in the setting of straddling and overriding of the tricuspid  
451 valve. The conduction axis in this setting (red bar) remains postero-inferior, but no longer arises  
452 from the regular atrioventricular node at the apex of the triangle of Koch (white star with red  
453 borders). Instead, the axis, carried on the malaligned ventricular septum, arises from an  
454 anomalous inferior atrioventricular node (red circle with white borders).

455

456



457

458 Figure 6. Incidence of various arterial trunk relationships observed within the archive,  
459 highlighting that, overall, an anterior and rightward aorta was the most commonly observed  
460 position.

461