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Transposition – Introduction

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In the past, most arguments concerning transposition devolved on how best to define it. When Mathew Baillie described the first case, at the end of the eighteenth century, he had no problems. He simply called it a singular malformation![1] Subsequent to Baillie's description, arguments raged amongst morphologists and pathologists as to whether the entity was best described on the basis of how the aorta was related to the pulmonary trunk, or according to the origin of the arterial trunks from inappropriate ventricles. There is little doubt now that all clinicians diagnose transposition on the basis of discordant origin of the arterial trunks from the ventricular mass, although there is still debate as to whether this discordant origin is best described in terms of connections[2] or alignments.[3] There remain some, nonetheless, who still describe entities such as "double outlet with transposition", using this phrase to describe the arrangement in which both arterial trunks arise from the right ventricle, but with the aorta positioned anteriorly. For those adopting the concepts of connections or alignments, this combination is clearly impossible.[4] Yet for those who choose to define "transposition" on the basis of the anterior location of the aorta,[5] this would remain an entirely logical description. These problems of nomenclature, and many more aspects of the morphology of discordant origin of the arterial trunks from the ventricular mass, are discussed at length in the first review of this part of the supplement,[6] co-authored by myself, who believes firmly in the concept of connections, and Paul Weinberg, who is equally convinced that the cardiac segments are best described in terms of alignments rather than connections. Our joint review hopefully shows that differences in nomenclature nowadays are relatively minor, and even where they exist, they do not stand in the way of reaching consensus on how best to describe the segmental combination in which the atrial chambers are joined to morphologically appropriate ventricle, but the ventricles support morphologically inappropriate arterial trunks. It is this particular morphological arrangement that, for the purposes of this section of our supplement, is described as "transposition".

In the other reviews within this part of the supplement, we concentrate on different surgical approaches and their sequels. Morrell et al.[7] discuss the options for surgical treatment of transposition when complicated by a ventricular septal defect and obstruction to the sub-pulmonary left ventricular outflow tract. The classical approach to this combination, of course, is the Rastelli procedure.[8] Morrell et al., however, discuss and illustrate the other options, including the widely quoted but poorly understood Rev procedure,[9] and the less widely quoted, but equally efficacious, surgical option promoted initially by Nikaidoh.[10] The preference of the group from St Petersburg is the Nikaidoh option, and their excellent results demonstrate the potential of this ingenious surgical procedure. Tchervenkov[11] then reviews the equally difficult combination of transposition with obstruction to the subaortic outflow tract, particularly when associated with severe coarctation or interruption of the aortic arch. In a wide-ranging review of the sequels of the arterial switch procedure, Mussatto and Wernovsky[12] then discuss not only the potential haemodynamic problems that can result from less than perfect surgical procedures, but focus attention on the psychosocial problems that can be the consequence of all operative procedures, at this point concentrating on the arterial switch. This review complements the wider ranging discussion of psychosocial aspects of surgical treatment of congenital cardiac disease presented by Mussatto later in our supplement.[13]

Prior to these essays on surgical procedures and their consequences, Lacour-Gayet and I discuss the anatomical and surgical aspects of perhaps the most

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important current morphological aspect of hearts with transposed arterial trunks, namely the arrangement of the coronary arteries.[14] Lacour-Gayet has based his surgical approach to coronary arterial transfer on the huge experience he gained whilst performing the arterial switch procedure during his appointment in Paris at Hôpital Marie-Lannelongue.[15] The surgeons working at this centre in Paris have now carried out the arterial switch procedure in more than 1,000 patients. François explains how, based on the concept of "looping" of the coronary arteries relative to the arterial pedicle, their chosen surgical technique for coronary arterial transfer is suitable for all variations in coronary arterial pattern. The problem remains, however, of how best to describe these variations. Paul Weinberg, in our joint chapter,[6] takes the stance that each individual case needs individual description, describing first the position of the aortic sinuses in space relative to the bodily coordinates, and then describing the how each of the three major coronary arteries, the right, circumflex, and anterior interventricular arteries, originates from the aortic valvar sinuses. No-one, least of all me, would seek to quarrel with this approach to description of the coronary arterial arrangement in the individual patient. When seeking to analyse large numbers of operative procedures so as to identify any anatomic risk factors for the arterial switch procedure in terms of coronary arterial anatomy, nonetheless, it is advantageous to have a simple means of describing the various arterial patterns. I remain convinced that this option is provided by the so-called "Leiden Convention".[16] Thus, in our joint review of the variations in coronary arterial anatomy, and its surgical significance, François Lacour-Gayet and I[14] show how the Leiden convention can be combined with the concept of "looping" so as to account for all variations that may be encountered by the surgeon transferring the coronary arteries in the setting of patients with transposition.

It is worth emphasising, in this context, that the Leiden convention[16] is no more than a means of comparing the origin of the coronary arteries from the aortic valvar sinuses in large numbers of cases, yet removing from the equation the additional variation of the interrelationships between the aorta and the pulmonary trunk. The convention works because, almost without exception, the coronary arteries, irrespective of the anatomic complexity to be found elsewhere in the heart, arise from one or other, or usually both, of the two aortic sinuses that are adjacent to the pulmonary trunk. These arterial sinuses, irrespective of the relationships of the arterial trunks, retain their adjacency to the pulmonary trunk (Fig. 1), even in the case of commissural mismatch.[17] Furthermore, the Leiden convention can readily be adapted, by recognising the location of the raphe, to account

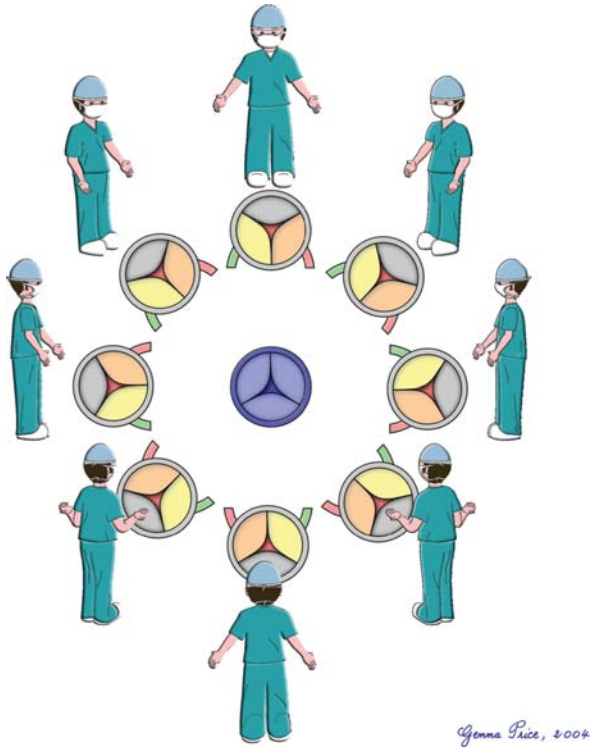

Figure 1.
The cartoon shows how, irrespective of the interrelationships of the aorta relative to the pulmonary trunk, the coronary arteries, almost without exception, take their origin from one or other, or usually both, of the aortic sinuses that are adjacent to the pulmonary trunk. The colour coding shows how the sinuses retain their position relative to the observer standing in the non-adjacent sinus irrespective of the relationships of the aorta to the pulmonary trunk. The colour coding for the sinuses, the main stem of the left coronary artery, and the three main coronary arterial branches, is the same as is used in the chapter by Lacour-Gayet and me[14] which appears in this part of the supplement.

for cases in which the coronary arteries arise from the same sinus in the setting of a bicuspid aortic valve. The key to the convention is to differentiate between the two aortic sinuses that, virtually without exception, give rise to the coronary arteries, since when seen relative to the coordinates of the body, these sinuses can occupy markedly different locations (Fig. 1). Differentiating the sinuses one from the other independent of their position in space can simply be done by viewing them from the stance of the non-adjacent aortic sinus. Irrespective of their position in space, one sinus is always to the right hand of the observer, whilst the other sinus is to the left hand. This approach works particularly well in the normal heart, since the right handed sinus gives rise to the right coronary artery, whilst the left handed sinus gives rise to the main stem of the left coronary artery (Fig. 2). A potential complication in the setting of transposition, found disturbing by some, is that, in the commonest

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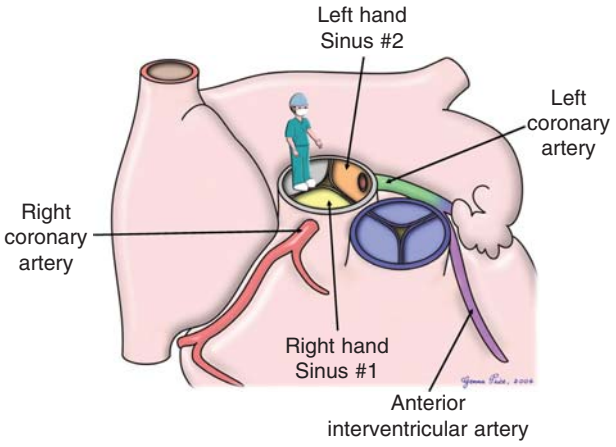

Figure 2.
In the normal heart, when viewed from the stance of the non-adjacent sinus of the aorta, the aortic sinus to the right hand of the observer gives rise to the right coronary artery, whilst the sinus to the left hand of the observer gives rise to the main stem of the left coronary artery. The colour coding is the same as used in the chapter by Lacour-Gayet and me.[14]

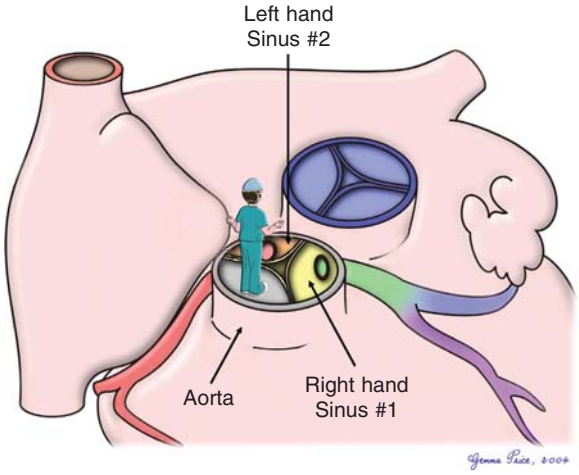

Figure 3.
In the commonest pattern of coronary arterial anatomy found in the setting of transposition, if viewed from the stance of the observer standing in the non-adjacent aortic sinus, the aortic sinus seen to the right hand of the observer, defined as "Sinus #1" in the original Leiden convention,[16] *gives rise to the main stem of the left coronary artery, whilst the right coronary artery arises from the sinus that is to the left hand of the observer. This sinus was designated as "#2" in the Leiden convention.*

variant of coronary arterial origin (Fig. 3), it is the aortic sinus to the right hand of the observer that gives rise to the main stem of the left coronary artery. This happening does not particularly bother me since, as I have explained, I see the major purpose of the Leiden convention as permitting the comparison of large numbers of cases without needing to worry about

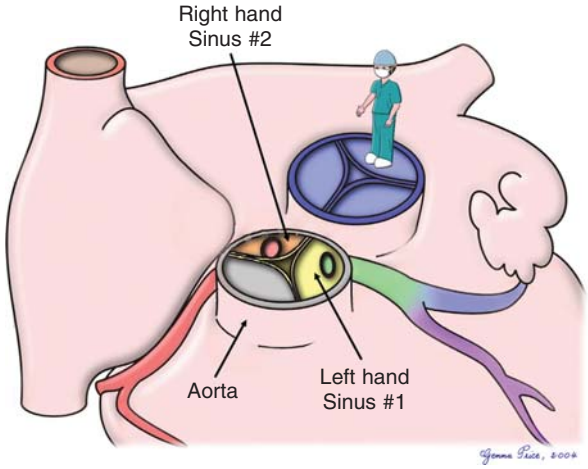

Figure 4.
If the observer, instead of standing in the non-adjacent aortic sinus, positions himself or herself in the non-adjacent pulmonary sinus, as suggested by Amato et al.,[19] *this sinus becoming the new aortic non-adjacent sinus after the arterial switch procedure, then the right coronary artery, in the commonest pattern seen in transposition, arises from the facing sinus to the right hand of the observer. This remains "Sinus #2", however, when named as suggested in the Leiden convention.*[14] *The main stem of the left coronary artery in this commonest pattern then arises from Sinus #1, this being the sinus to the left hand of the observer when viewed from the non-adjacent sinus of the pulmonary trunk.*

the specific relationship of the aorta relative to the pulmonary trunk. The huge complexity of the categorisation devised by Shaher and Puddu[18] serves to emphasise the difficulties that arise when attempts are made to describe coronary arterial anatomy in a system that also takes into account the relationship of the aorta to the pulmonary trunk. The potential problem of the right handed sinus giving rise to the main stem of the left coronary artery, however, can be overcome simply by viewing the aortic root from the stance of the non-adjacent sinus of the pulmonary trunk.[19] This approach (Fig. 4) has particular surgical utility because the pulmonary root becomes part of the new aorta once the arterial switch has successfully been accomplished. Even using this approach, the situation will still arise in which the right handed sinus will give rise to the main stem of the left coronary artery. Such instances, however, will be relatively infrequent, and perhaps serve to emphasise that they are, indeed, unusual variants. It also remains a fact, nonetheless, that most surgeons circumvent the problems of distinguishing right handed or left handed sinuses simply by following the original precedent of the team from Leiden,[16] and describing the sinus seen to the right hand, if viewing the root from the aorta, or to the left hand if taking a position in the pulmonary trunk, as "Sinus #1". "Sinus #2" is obviously the complementary adjacent sinus of the aortic root.

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Although generally having an aversion to numeric categorisations, and still having to think hard to remember which is "Sinus #1" in terms of handedness, it seems to me that this solution provides the best means of differentiating between the aortic sinuses irrespective of their location in space relative to the coordinates of the body. And, despite recognising the validity of describing individual cases in terms of bodily coordinates, I retain my belief that the Leiden convention[16] is the best means of comparing the coronary arterial anatomy in large numbers of patients with transposition.

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