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The nomenclature, definition and classification of discordant atrioventricular connections

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CONGENITALLY CORRECTED TRANSPOSITION IS A complex cardiac lesion that is often associated with ventricular septal defect, obstruction of the outflow tract of the morphologically left ventricle, and abnormalities of the morphologically tricuspid valve.[1,2] Nomenclature for this lesion has been variable and confusing.[1] In this review, we define, and hopefully clarify this terminology. The lesion is a combination of discordant union of the atrial chambers with the ventricles, and the ventricles with the arterial trunks.[1,2] In rare circumstances, discordant atrioventricular connections can be associated with concordant ventriculo-arterial connections. This malformation has been called "isolated ventricular inversion". The term is less than precise, and the descriptive approach using the phrase "*discordant atrioventricular connections with concordant ventriculo-arterial connections*" is preferred, as discussed below.

In 2000, Wilkinson, Cochrane, and Karl, on behalf of the International Congenital Heart Surgery Nomenclature and Database Project, proposed a definition and provided a classification for discordant atrioventricular connections.[1] The topic had been the subject of extensive debates and review during the meetings of members of the Society of Thoracic Surgeons and the European Association for Cardio-Thoracic Surgery.[1] Efforts were made to include all relevant categories of nomenclature, using synonyms where appropriate. The topic was further debated at the fifth meeting of the Nomenclature Working

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Group, which was held in the Mazurian Lake District, Poland, over the period July 19–23, 2004. It has been the task of the Nomenclature Working Group to establish the International Pediatric and Congenital Cardiac Code. This goal has been achieved by reviewing 28 major categories of cardiac lesions, and crossmapping several existing systems for nomenclature, including that prepared on behalf of the European Association for Cardio-Thoracic Surgery and the Society of Thoracic Surgeons, and the alternate version prepared for the Association for European Paediatric Cardiology.[3–8] This International Pediatric and Congenital Cardiac Code was unveiled officially during the Second International Summit on Nomenclature for Paediatric and Congenital Heart Disease, held at the Fourth World Congress of Pediatric Cardiology and Cardiac Surgery in Buenos Aires, Argentina, on September 19, 2005 [www.ipccc.net].

In this review, we will refer to the International Paediatric and Congenital Cardiac Code as the International Code, and we will present two versions of this International Code for diagnoses and procedures related to congenitally corrected transposition:

- The version derived from the International Congenital Heart Surgery Nomenclature and Database Project of the European Association for Cardio-Thoracic Surgery and the Society of Thoracic Surgeons.
- The version derived from the European Pediatric Cardiac Code of the Association for European Pediatric Cardiology.

These two systems were developed with considerable input from both cardiologists and surgeons. It is the crossmap developed for diagnoses and procedures related to discordant atrioventricular connections and related malformations that will be the focus of this review, following the steps taken previously in regard to the functionally univentricular heart[9] and hypoplastic left heart syndrome.[10] The current imperative of the group is to provide a classification of cardiac phenotypes, and not genotypes. In the future, knowledge of the underlying genetic defects may modify the schemes currently developed for classification.

Definitions related to discordant atrioventricular connections

"*Congenitally corrected transposition*" describes the lesion in which the morphologically right atrium is connected to the morphologically left ventricle, and the morphologically left atrium to the morphologically right ventricle. The aorta then takes its origin from the morphologically right ventricle, and the pulmonary trunk from the morphologically left ventricle. As a result of the discordant connections, systemic venous blood continues to flow to the pulmonary circulation, and pulmonary venous return passes to the systemic circulation, so that the circulations are physiologically corrected.

The malformation may occur in patients with the normal atrial arrangement, otherwise known as "situs solitus" where solitus denotes usual, or in those with mirror-imaged atrial arrangement, otherwise known as "situs inversus". It cannot occur in the setting of visceral heterotaxy, when the atrial appendages are isomeric, because it is impossible for the atrial and ventricular chambers to be joined in discordant fashion when both appendages have the same morphology. Almost always the venoatrial connections are grossly abnormal in the setting of heterotaxy, but rarely the flows of blood can mimic the arrangement seen in corrected transposition.

The term corrected transposition was first used by Rokitansky.[11] By 1956, Cardell[12] was able to review 25 cases, including his own, and in 1961, Lev and Rowlatt described several autopsied specimens.[13] As far as we know, the term congenitally corrected transposition was introduced by Schiebler et al. in 1961,[14] seeking to distinguish surgically repaired transposition with concordant atrioventricular connections from the congenitally corrected variant. While the term "corrected transposition" has gained widespread use, it has been opposed by some cardiac morphologists, and is not the favored nomenclature of others.[13,15,16] In this respect, the term "transposition" itself has been controversial. Some early workers used the term to define any abnormality in aorto-pulmonary relationships.[11] More usually, those using the term required antero-posterior reversal of the aorto-pulmonary spatial relationship, along with the presence of a muscular subaortic conus.[12,17,18] In 1971, Van Praagh et al. proposed the straightforward, and currently favoured, definition of transposition: "right ventricular origin of the aorta and left ventricular origin of the pulmonary artery".[19] The term "transposition", therefore, is synonymous with origin of the arterial trunks from morphologically inappropriate ventricles,[20,21] so there seems little reason for arguing with the congenital correction of such transposition when the atrial chambers are similarly joined to morphologically inappropriate ventricles.

On this basis, therefore, the Nomenclature Working Group offers the following definition for the term "congenitally corrected transposition":

"Congenitally corrected transposition is synonymous with the terms 'corrected transposition' and 'discordant atrioventricular connections with discordant ventriculo-arterial connections', and is defined as a spectrum of cardiac malformations

where the atrial chambers are joined to morphologically inappropriate ventricles, and the ventricles then support morphologically inappropriate arterial trunks."

Although congenitally corrected transposition can occur in isolation, it is usually associated with additional anomalies, including one or more ventricular septal defects, obstruction of the outflow tract of the morphologically left ventricle, abnormalities of the morphologically tricuspid valve,[22] or abnormalities of atrioventricular conduction, which may also evolve and progress as part of the natural history.[2] Although incompetence of the morphologically tricuspid valve is less frequent than in Ebstein's malformation in patients with concordant atrioventricular connections,[14] severe tricuspid incompetence is probably the single most important adverse prognostic feature of congenitally corrected transposition.[23] Rarely, there may be co-existent pulmonary atresia or hypoplasia of the morphologically left ventricle, and even more rarely, hypoplasia of the morphologically right ventricle.

Over time, various terminologies have been used to describe "congenitally corrected transposition" and related malformations. We will discuss briefly some of these terms.

Discordant atrioventricular connections with discordant ventriculo-arterial connections: This combination represents the recommended name for corrected transposition. The terms "corrected transposition", "congenitally corrected transposition", and "discordant atrioventricular connections with discordant ventriculo-arterial connections" are all acceptable synonyms.

Double discordance: This term is not uniformly used, although it does have the virtues of brevity and accuracy. It is felt by some to be an acceptable synonym for congenitally corrected transposition. Others criticize the term for a potential lack of specificity, because it does not clearly state what is discordant. This conflict demonstrates why it is best to employ segmental nomenclature, and specify "discordant atrioventricular connections with discordant ventriculo-arterial connections".

Discordant atrioventricular connections (with transposition): The term "discordant atrioventricular connections" has been employed as an alternative to "corrected transposition", both with and without the qualifying phrase "with transposition." When used in isolation, it is clearly inaccurate, and can be a cause of confusion and imprecision. If qualified by such phrases as "with discordant ventriculo-arterial connections" or "with transposition", it may be regarded as accurate and explicit.[1]

Discordant transposition: This term is favoured by some as a shorthand term for the malformation. In practice, it is no briefer than the more widely used "corrected transposition". Using Boolean logic, the term discordant transposition is intended to imply combined discordant atrioventricular and ventriculo-arterial connections, and in that sense is precise and useful, being the preferred term of surgeons and cardiologists working at the University of California-San Francisco, among others.[1] Others criticize this term because the word "discordant" is an adjective, and the only word in this name that it could possibly modify is transposition. Since transposition is the term for discordant ventriculo-arterial connections, the meaning of this phrase is literally "discordant discordant ventriculo-arterial connections", which logically implies concordant ventriculo-arterial connections. This criticism demonstrates why it is best to employ segmental approach and specify "discordant atrioventricular connections with discordant ventriculo-arterial connections".

Ventricular inversion: The term "inversion" has been used widely to describe left-right reversal, or mirror-imagery, of paired structures.[14] When used to describe the ventricular mass, this term is therefore synonymous with l-looping, and normally implies that the morphologically left ventricle is right sided, and the morphologically right ventricle is left sided. Although ventricular inversion is most commonly seen in association with the combination of normal atrial position and transposition, this is not always the case, and the term "ventricular inversion" does not specify the nature of the atrioventricular or ventriculo-arterial connections. Unfortunately, the term "ventricular inversion", has also been used to describe hearts in which the ventricles are normally related, with d-loop ventricles, in patients with mirror-imaged atrial arrangement, or "situs inversus",[24] arguing that it is synonymous with the more widely understood term "discordant atrioventricular connections".[21,25] This usage of the term "ventricular inversion" is highly confusing, and should be avoided. The term "isolated ventricular inversion" was originally introduced to imply ventricular inversion or l-looping with normal atrial situs and without transposition, which in segmental shorthand is {S,L,S}, [20] and the term "isolated ventricular non-inversion" was introduced to specify hearts with mirror-imaged atrial arrangement, or atrial situs inversus, d-loop ventricles, and inverted normally related great arteries {I,D,I}. These terms were therefore intended to specify discordant atrioventricular connections with concordant ventriculoarterial connections, resulting in circulations that are physiologically uncorrected. The terms have not been universally used in this fashion, and their meaning is inherently ambiguous because the sidedness of the great arteries does not specify whether the ventriculo-arterial connections are concordant or discordant. Because of

this confusing situation, the terms "isolated ventricular inversion", and "isolated ventricular non-inversion", should be avoided.

Malposition of the great arteries: Malposition of the great arteries is a term that defines or includes all defects with abnormal position of the great vessels, regardless of the ventricular origin.[26] The distinction from "transposition" is important, albeit elusive. As discussed above, transposition should now be considered synonymous with "discordant ventriculo-arterial connections",[1,20,21] and although transposition is a form of malposition, malposition can occur in the setting of either concordant or discordant ventriculoarterial connections. Other forms of "malposition" include double-outlet right ventricle, double-outlet left ventricle, and the so-called anatomically corrected malposition with its variants.[19,27] It is beyond the scope of our review to discuss these niceties.

l-transposition ("L-TGA"), d-transposition ("D-TGA"), and Complete transposition: In the 1960s, the term "l-transposition" or "L-TGA" was introduced by Van Praagh[16] to describe hearts with a left-sided, and usually anterior, aorta arising from the morphologically right ventricle, the pulmonary trunk taking its origin from the other ventricle. In most cases, this arrangement coexisted with discordant atrioventricular connections when the morphologically left ventricle was right sided, and the morphologically right ventricle left sided. Van Praagh designated this ventricular arrangement as an "l-loop".[16] In most cases, l-transposition is indicative of corrected transposition. It is well recognized, nonetheless, that some patients with corrected transposition, even in the presence of an l-loop, have an aorta that lies anterior and to the right of the pulmonary trunk, a situation which may be denoted by the notation {S,L,D}. This breaches the "loop rule", as has also been noted in patients with transposition and concordant atrioventricular connections.[28] Most patients with corrected transposition in the setting of mirror-imaged atrial arrangement, or atrial situs inversus, however, exhibit a d-loop and d-transposition, a situation which may be denoted by the notation {I,D,D}.[1,27] For these reasons, "l-transposition" is obviously unsuitable for use as a substitute for "corrected transposition", and usage in this fashion is imprecise, and should be discouraged.[1,27] The terms "d-transposition", "a-transposition" and "l-transposition" define no more than the spatial arrangements of the discordantly connected arterial trunks. The term "d-transposition", therefore, refers only to hearts with transposition in which the aortic valve is to the right of the pulmonary valve. This anatomic arrangement is the most common variant of transposition with concordant atrioventricular connections, being found, according to Jaggers et al.,[27] in more than four-fifths of cases reported by Van Praagh. The terms complete transposition, and incomplete transposition, are obsolete. In the past, it was double-outlet right ventricle that was termed incomplete transposition. Hence, complete transposition may refer to either physiologically uncorrected or corrected transposition, because in both cases the arterial trunks arise from morphologically inappropriate ventricles, and thus are "completely transposed". When used as a modifier of transposition, therefore, the term "complete" is redundant, and should be avoided.[27] Although most patients with physiologically uncorrected transposition have anterior and right-sided aortas, not all patients with d-transposition are physiologically uncorrected, and not all patients with physiologically uncorrected transposition have d-transposition.[27] Physiologically uncorrected transposition includes hearts with the segmental anatomy {S,D,D}, {S,D,A}, and {S,D,L}, as well as {I,L,L} and {I,L,D}. Similarly, the term "l-transposition" is often used erroneously as a synonym for corrected transposition. But not all cases of l-transposition are corrected transposition,[27] while corrected transposition includes hearts with the segmental anatomy {S,L,L}, {S,L,D}, and {I,D,D}.

The terms "d-transposition", "a-transposition" and "l-transposition" cannot, therefore, be used to imply or define the presence of corrected transposition or uncorrected transposition. These terms merely define the spatial arrangements of the discordantly connected arterial trunks and do not specify whether the atrioventricular connection is concordant or discordant. They are, nonetheless, important modifiers because the spatial relations of the great vessels may closely correlate with the anatomic features of the coronary arteries.[27]

Regardless of the original intent of these terms, many people use "d-transposition" to refer to the combination of d-looped ventricles and transposition, and "l-transposition" to imply the combination of l-looped ventricles and transposition, without reference to the position of the arterial roots. In this use, {S,D,L} transposition is a form of d-transposition, and {I,L,D} is form of l-transposition. Using this interpretation, the terms "d-transposition" and "l-transposition" refer to d-loop transposition and l-loop transposition. This usage adds even more confusion, because it is apparent that universal agreement does not exist as to whether or not the "d" and "l" refer to arterial position or ventricular looping. This point alone potentially represents the worst problem with the terms "d-transposition" and "l-transposition", and further justifies the recommendation to avoid these terms.

Physiologically corrected transposition: In congenitally corrected transposition, as a result of the double discordance, the defect is "physiologically corrected". In clinical practice, however, corrected transposition often is far from physiologically correct because of the associated defects, which are the rule rather than

the exception. The term "physiologically corrected transposition" is therefore of little practical use.[1]

In the past, the rare situation in which the aorta arises from the morphologically left ventricle in an anterior position, with the pulmonary trunk from the morphologically right ventricle lying posteriorly, was described as anatomically corrected transposition. In some of the patients with this arrangement, the atrioventricular connections may be discordant, so that the physiology is similar to that of physiologically uncorrected transposition. This lesion is now termed "anatomically corrected malposition", albeit that some variants with spiralling rather than parallel arterial trunks can still present problems in nomenclature. Such details are beyond our scope in this review. Fortunately, the currently favoured definition of transposition, being synonymous with discordant ventriculo-arterial connections, solves the dilemma of "anatomically corrected malposition", since obviously hearts with the arterial trunks arising from appropriate ventricles can no longer be considered to show "transposition".

Mixed levocardia and dextrocardia: These terms were introduced by Lev,[29] and are not in current use. Mixed levocardia referred to the situation in which the apex of the heart pointed to the left, but in which the arrangement of atriums and ventricles was not as expected.[13] The related term "mixed dextrocardia"[29] catered similarly for patients with discordant atrioventricular connections and right-sided hearts and apexes. The terms are generally not used nor understood, and are not recommended. The term "dextroversion" is similarly no substitute for corrected transposition. It refers to hearts in patients with the normal atrial arrangement and rightward displacement or rotation of the ventricles.[30–32] In other words, the terms "dextrocardia" and "dextroversion" are not synonyms for corrected transposition; dextrocardia is synonymous with a right-sided ventricular mass and dextroversion is synonymous with a right-sided ventricular apex.

Inverted transposition: This term has similar meaning to Van Praagh's term "l-transposition".[13,15] It has not been widely used, and is best avoided.

Alignments and connections: The essence of the original segmental approach to diagnosis and nomenclature was analysis of the topological arrangement of the atrial chambers, the ventricular mass, and the arterial trunks.[16] When the European school[21] sought to develop further this innovative methodology, they emphasized the importance also of describing the way the basic segments were joined together across their junctions. The Europeans described these variations in terms of connections.[33] It was unfortunate, therefore, that in their original account they chose the terms "concordance" and "discordance" to describe the appropriate and inappropriate junctional connections. This choice of terminology was less than perfect, since the Bostonian school had used "concordance" and "discordance" to describe segmental harmony or disharmony, irrespective of the precise connections across the segmental junctions.[34] For example, in the Boston terminology all {S,D,D} hearts have ventriculoarterial concordance and all {S,D,L} hearts have ventriculoarterial discordance, regardless of whether the left ventricle connects to the aorta or the pulmonary artery. In other words, the Boston school defines concordance or discordance between segments according to segmental harmony or disharmony, that is the relationship between atrial situs (S) or (I), ventricular loop (D) or (L), and spatial arterial relations (S) or (L),[34] irrespective of the precise way the segments joined one another across the junctions. The original European terminology used the term "ventriculoarterial concordance" to describe those hearts in which the left ventricle connected to the aorta regardless of the segmental arrangements. Recognising their own mistake, when revising their approach, the Europeans pointed out that the ambiguity in terminology could be resolved by describing specifically concordant or discordant atrioventricular and ventriculo-arterial connections. The European school, therefore, modified their own nomenclature, arguing that "concordance" or "discordance" should no longer be used as nouns, but that "concordant" and "discordant" should be used as adjectives to qualify both the atrioventricular and ventriculo-arterial connections.[33] The Europeans further emphasized that, for the purposes of defining these connections, the ventricular mass was defined as extending from the atrioventricular to the ventriculo-arterial junctions.

The term alignment was subsequently introduced by Van Praagh et al.[35] with the argument that atriums and ventricles, as well as the ventricles and great arteries, do not connect to each other at all but, instead, are separated by intermediate "connecting segments", namely the atrioventricular canal and the conuses. In this context, they[35] have described two additional segments, and consequently defined atrioventricular alignments and ventriculo-arterial alignments as concordant or discordant. The Bostonian approach, therefore, was to describe alignments of the segments, rather than connections between them.[36]

The European school identifies three problems with these suggestions. First, it is unclear how the "atrioventricular canal" is to be distinguished in the postnatal heart. Second, if the ventricular mass is defined as extending from the atrioventricular to the ventriculo-arterial junctions, then the conuses are an integral part of this ventricular mass. Third, there is no way of distinguishing in their own right the

conuses which support the aorta as opposed to the pulmonary trunk.

Very rarely, the segmental topological arrangements do not correspond with the way the chambers and arterial trunks are joined across their junctions.[37] It is essential, therefore, that any system of nomenclature is able to distinguish between segmental topologies and junctional variations.[37] This goal may be accomplished either via the European approach or the Bostonian approach if care is taken to specify both these features. Consider the rare heart that would be described in the European school as showing discordant atrioventricular and ventriculo-arterial connections in the setting of usual atrial arrangement with right hand ventricular topology and anterior and right sided aorta. With the Bostonian approach, this heart could be described as congenitally corrected transposition {S,D,D}. Such hearts exemplify the principle that it is essential to describe both segmental topologies and junctional connections, a point that is again emphasized in the discussion of the criss-cross atrioventricular valve below.

{S,L,L}, {S,L,D}, and {I,D,D}: Although the terms l-transposition or "L-TGA", as well as d-transposition or "D-TGA", are best avoided, because of the reasons discussed above, a complete segmental description is acceptable when describing hearts with congenitally corrected transposition. As also discussed above, corrected transposition includes hearts with the segmental anatomy {S,L,L}, {S,L,D}, and {I,D,D}. The segmental combinations in isolation, however, are not synonymous with corrected transposition. This is because double outlet from either the right or left ventricles can be described in this fashion when associated with discordant atrioventricular connections. Furthermore, in exceedingly rare circumstances, again as discussed above, hearts with discordant atrioventricular connections can exhibit disharmony in terms of the segmental connections. In these rare circumstances[38,39] the usually positioned right atrium is joined to the abnormally positioned morphologically left ventricle even though there is right hand ventricular topology. In this rare circumstance, therefore, the segmental notation {S,D,*}, or {I,L,*} in the even rarer setting of mirror-imagery, would not describe the presence of the discordant atrioventricular connections.

Criss-cross atrioventricular valves and upstairs-downstairs ventricular relationships: The two atrioventricular inlets are usually aligned more or less in parallel, each being at approximately the same level in the thorax. Under such circumstances, the ventricles are related in the anticipated manner, with one being to the right of the other, although sometimes one ventricle is more anterior or superior. Rarely, one atrioventricular valve and inlet may reside at a significantly higher level than its counterpart, and in extreme cases, the tricuspid valve and right ventricular inlet may be directly anterosuperior to the mitral valve and left ventricular inlet. If the ventricular mass is simply tilted along its long axis, the ventricles may be in a superoinferior relationship, sometimes termed "upstairs-downstairs". This term "upstairs-downstairs" might be best avoided, as one would not think of other inferior-superior structures, such as the two caval veins, as "upstairs-downstairs". If the ventricular mass is rotated, or twisted around its long axis such that the axes of the atrioventricular inlets cross one another in the transverse plane, an atrioventricular "criss-cross" relationship is present.[40,41] The majority of hearts with criss-cross atrioventricular relationships have concordant atrioventricular alignments and connections. Specifically, almost all have the usual atrial arrangement (atrial situs solitus) and d-ventricular loop.[38] This "criss-cross" relationship may also be seen, nonetheless, with discordant atrioventricular connections. In this situation, despite discordant atrioventricular connections with usual atrial arrangement or situs solitus, the morphologically right ventricle may lie on the right side of the ventricular mass, with the left ventricle being inferior and leftward, forming the left-sided cardiac apex. Even in such rare cases, it should still be possible to determine the topology of the ventricular mass, which can be disharmonious with the expected atrioventricular connections. In such cases, as emphasized above, it is essential to describe both the connections and the ventricular topology.

Discordant atrioventricular connections with concordant ventriculo-arterial connections: Unlike corrected transposition, the physiology of discordant atrioventricular connections with concordant ventriculo-arterial connections resembles that of transposition, with deoxygenated blood passing to the systemic circulation via the morphologically left ventricle which is connected to the aorta, and oxygenated blood being channeled to the pulmonary trunk from the morphologically right ventricle. This defect was sometimes called "isolated ventricular inversion" and is extremely uncommon. As we have already discussed, it is best to avoid the potentially confusing name "isolated ventricular inversion", and instead use the specific terminology "discordant atrioventricular connections with concordant ventriculo-arterial connections". As is always the case with very rare defects, the best strategy is to use the segmental approach, describing atrial arrangement, atrioventricular connections, ventriculo-arterial connections, and other aspects including the spatial relationships of the various structures and associated malformations in simple anatomic terms.[1] The abbreviated segmental formula is of limited help in these complex situations, though in many instances with discordant

atrioventricular and concordant ventriculo-arterial connections, the arterial relationship is either normal when the ventricles are normally related, or mirror-imaged with respect to normal when the ventricles are in the mirror-imaged relationship. The segmental formulas for such relationships are: "D" (d-loop) with "S" (solitus) arterial relationship, with solitus equaling the usual arterial relationship; and "L" (l-loop) with "I" (inversus) arterial relationship, inversus then indicating the mirror-imaged arterial relationship. Of note, in most instances of discordant atrioventricular and concordant ventriculo-arterial connections, the arterial trunks are parallel and not spiralling. Thus, with discordant atrioventricular and concordant ventriculo-arterial connections, the complete segmental formula is usually {S,L,D} or {I,D,L} and less commonly {S,L,I} or {I,D,S}. The arguments as to what is "normally related" in these settings is beyond the scope of this review, but is an important consideration. The important point to reemphasize is that it is essential to describe separately the connections, the atrial arrangement, the ventricular topology, and the spatial arrangements of the arterial trunks.

Discordant atrioventricular connections with double-outlet right ventricle

This is a rare malformation, which frequently resembles corrected transposition with ventricular septal defect.[1] The associated interventricular connection is typically in subpulmonary position. The physiology, therefore, is similar to corrected transposition with ventricular septal defect, as is the surgical treatment. As with other forms of double outlet right ventricle, nonetheless, there is a wide variety of associated malformations and variations, which often profoundly affect the clinical presentation and management.

Corrected transposition

The term "corrected transposition" is widely used, and generally well understood by pediatric cardiologists and surgeons. Phrases such as "discordant atrioventricular connections and discordant ventriculo-arterial connections" and "discordant atrioventricular connections with transposition", are very acceptable, accurate, and explicit alternatives. "Congenitally corrected transposition" is also very widely used, and is well understood by adult cardiologists and surgeons who do not deal with this lesion very often.[1] Segmental nomenclature such as {S,L,L} and {I,D,D} can be used as an adjunct to "corrected transposition", but neither should be used in isolation. The use of the abbreviated segmental designation "l-transposition" to refer to corrected transposition is less satisfactory and should be discouraged.

Therapy for discordant atrioventricular connections

Operations for congenitally corrected transposition fall basically into five categories:

- Palliative procedures
- Definitive operations that leave the morphologically right ventricle in the systemic circulation and the morphologically left ventricle in the pulmonary circulation, also known as "classic repairs"[42-45]
- Definitive operations that leave the morphologically left ventricle in the systemic circuit and the morphologically right ventricle in the pulmonary circuit, also known as "anatomic repairs". These include the "*atrial switch and arterial switch*", also termed the "*double switch*",[43,44,46] *the "atrial switch and Rastelli procedure*",[46,47] the "*atrial switch and intraventricular tunnel repair with reversed tunnel of Patrick and McGoon*",[46] and the "*atrial switch and Nikaidoh procedure*", which includes translocation of the aortic root into the morphologically left ventricle combined with the atrial switch"[48]
- Definitive operations that create a one-and-one half ventricular repair, such as closure of a ventricular septal defect combined with a superior cavopulmonary anastomosis[45]
- Definitive operations that leave both ventricles connected to the systemic circuit and that employ some type of cavopulmonary connection for the systemic venous to pulmonary arterial circuit, in other words, Fontan-type procedures[48]

Many of the palliative procedures used in patients with corrected transposition have broad applicability in surgery for other types of complex cardiac lesions. These palliative procedures, such as systemic-to-pulmonary shunts and banding of the pulmonary trunk, will not be discussed further, because they can be applied to any of a variety of congenital cardiac malformations. They are covered in separate sections of the overall lists dealing with palliative procedures. The biventricular repairs, in contrast, are in many aspects unique to congenitally corrected transposition.

The classic repair for congenitally corrected transposition with ventricular septal defect is no more than simple closure of the defect, leaving the morphologically right ventricle to pump the systemic circuit. Although the initial outcome of such repairs has been generally good in properly selected patients, at 10 years the results can be disappointing, primarily due to problems with function of the morphologically tricuspid valve and right ventricle.[42–44,46] For this reason, there has been great interest in alternate strategies, with the goal of creating an "anatomic repair". While much more complex, such operations, as listed above, may have a better long-term result.

Table 1. The European Association for Cardiothoracic Surgery – Society of Thoracic Surgeons derived version of the International Pediatric and Congenital Cardiac Code for Diagnoses related to Discordant Atrioventricular Connections.

Discordant AV connections with situs inversus	01.04.01, 01.03.01
Discordant AV connections with situs inversus, Concordant VA connection ("Isolated ventricular inversion") (usually {IDL} or {IDS})	01.04.01, 01.03.01, 01.05.00
Discordant AV connections with situs inversus, Concordant VA connection ("Isolated ventricular inversion") (usually {IDL} or {IDS}), IVS	01.04.01, 01.03.01, 01.05.00, 07.21.00
Discordant AV connections with situs inversus, Concordant VA connection ("Isolated ventricular inversion") (usually {IDL} or {IDS}), VSD	01.04.01, 01.03.01, 01.05.00, 07.10.00
Discordant AV connections with situs inversus, Crisscross AV connection and discordant VA connection (Corrected transposition/crisscross)	01.01.03, 01.03.01, 02.03.03
Discordant AV connections with situs inversus, Crisscross AV connection and discordant VA connection (Corrected transposition/crisscross), IVS and LVOTO	01.01.03, 01.03.01, 02.03.03, 07.21.00, 07.09.01
Discordant AV connections with situs inversus, Crisscross AV connection and discordant VA connection (Corrected transposition/crisscross), IVS and no LVOTO	01.01.03, 01.03.01, 02.03.03, 07.21.00, 07.09.01+ Q1.90.81
Discordant AV connections with situs inversus, Crisscross AV connection and discordant VA connection (Corrected transposition/crisscross), VSD and LVOTO	01.01.03, 01.03.01, 02.03.03, 07.10.00, 07.09.01
Discordant AV connections with situs inversus, Crisscross AV connection and discordant VA connection (Corrected transposition/crisscross), VSD and no LVOTO	01.01.03, 01.03.01, 02.03.03, 07.10.00, 07.09.01+ Q1.90.81
Discordant AV connections with situs inversus, Discordant VA connection (Congenitally corrected transposition) (usually {IDD})	01.01.03, 01.03.01
Discordant AV connections with situs inversus, Discordant VA connection (Congenitally corrected transposition) (usually {IDD}), IVS and LVOTO	01.01.03, 01.03.01, 07.21.00, 07.09.01
Discordant AV connections with situs inversus, Discordant VA connection (Congenitally corrected transposition) (usually {IDD}), IVS and no LVOTO	01.01.03, 01.03.01, 07.21.00, 07.09.01+ Q1.90.81
Discordant AV connections with situs inversus, Discordant VA connection (Congenitally corrected transposition) (usually {IDD}), VSD and LVOTO	01.01.03, 01.03.01, 07.10.00, 07.09.01
Discordant AV connections with situs inversus, Discordant VA connection (Congenitally corrected transposition) (usually {IDD}), VSD and no LVOTO	01.01.03, 01.03.01, 07.10.00, 07.09.01+ Q1.90.81
Discordant AV connections with situs inversus, Double-outlet right ventricle	01.04.01, 01.03.01, 01.01.04
Discordant AV connections with situs inversus, Double-outlet right ventricle, Aorta anterior to pulmonary artery ({IDA})	01.04.01, 01.03.01, 01.01.04, 02.06.03
Discordant AV connections with situs inversus, Double-outlet right ventricle, Aorta to left (or left anterior) of pulmonary artery ({IDL})	01.04.01, 01.03.01, 01.01.04, 02.06.04
Discordant AV connections with situs inversus, Double-outlet right ventricle, Aorta to right (or right anterior) of pulmonary artery ({IDD})	01.04.01, 01.03.01, 01.01.04, 02.06.02
Discordant AV connections with situs solitus	01.04.01, 01.03.00
Discordant AV connections with situs solitus, Concordant VA connection ("Isolated ventricular inversion") (usually {SLD} or {SLI})	01.04.01, 01.03.00, 01.05.00
Discordant AV connections with situs solitus, Concordant VA connection ("Isolated ventricular inversion") (usually {SLD} or {SLI}), IVS	01.04.01, 01.03.00, 01.05.00, 07.21.00
Discordant AV connections with situs solitus, Concordant VA connection ("Isolated ventricular inversion") (usually {SLD} or {SLI}), VSD	01.04.01, 01.03.00, 01.05.00, 07.10.00
Discordant AV connections with situs solitus, Crisscross AV connection and discordant VA connection (Corrected transposition/crisscross)	01.01.03, 01.03.00, 02.03.03
Discordant AV connections with situs solitus, Crisscross AV connection and discordant VA connection (Corrected transposition/crisscross), IVS and LVOTO	01.01.03, 01.03.00, 02.03.03, 07.21.00, 07.09.01
Discordant AV connections with situs solitus, Crisscross AV connection and discordant VA connection (Corrected transposition/crisscross), IVS and no LVOTO	01.01.03, 01.03.00, 02.03.03, 07.21.00, 07.09.01+ Q1.90.81
Discordant AV connections with situs solitus, Crisscross AV connection and discordant VA connection (Corrected transposition/crisscross), VSD and LVOTO	01.01.03, 01.03.00, 02.03.03, 07.10.00, 07.09.01
Discordant AV connections with situs solitus, Crisscross AV connection and discordant VA connection (Corrected transposition/crisscross), VSD and no LVOTO	01.01.03, 01.03.00, 02.03.03, 07.10.00, 07.09.01+ Q1.90.81
Discordant AV connections with situs solitus, Discordant VA connection (Congenitally corrected transposition) (usually {SLL})	01.01.03, 01.03.00
Discordant AV connections with situs solitus, Discordant VA connection (Congenitally corrected transposition) (usually {SLL}), IVS and LVOTO	01.01.03, 01.03.00, 07.21.00, 07.09.01
Discordant AV connections with situs solitus, Discordant VA connection (Congenitally corrected transposition) (usually {SLL}), IVS and no LVOTO	01.01.03, 01.03.00, 07.21.00, 07.09.01+ Q1.90.81
Discordant AV connections with situs solitus, Discordant VA connection (Congenitally corrected transposition) (usually {SLL}), VSD and LVOTO	01.01.03, 01.03.00, 07.10.00, 07.09.01

(*Continued*)

Table 1. (*Continued*)

Discordant AV connections with situs solitus, Discordant VA connection (Congenitally corrected transposition) (usually {SLL}), VSD and no LVOTO	01.01.03, 01.03.00, 07.10.00, 07.09.01+ Q1.90.81
Discordant AV connections with situs solitus, Double-outlet right ventricle	01.04.01, 01.03.00, 01.01.04
Discordant AV connections with situs solitus, Double-outlet right ventricle, Aorta anterior to pulmonary artery ({SLA})	01.04.01, 01.03.00, 01.01.04, 02.06.03
Discordant AV connections with situs solitus, Double-outlet right ventricle, Aorta to left (or left anterior) of pulmonary artery ({SLL})	01.04.01, 01.03.00, 01.01.04, 02.06.04
Discordant AV connections with situs solitus, Double-outlet right ventricle, Aorta to right (or right anterior) of pulmonary artery ({SLD})	01.04.01, 01.03.00, 01.01.04, 02.06.02
Discordant AV connections-modifier, AV valve, Overriding	06.00.01
Discordant AV connections-modifier, AV valve, Overriding, Left sided AV Valve	06.00.03
Discordant AV connections-modifier, AV valve, Overriding, Mitral valve	06.02.05
Discordant AV connections-modifier, AV valve, Overriding, Right sided AV Valve	06.00.02
Discordant AV connections-modifier, AV valve, Overriding, Tricuspid valve	06.01.05
Discordant AV connections-modifier, AV valve, Straddling	06.00.04
Discordant AV connections-modifier, AV valve, Straddling, Left sided AV Valve	06.00.06
Discordant AV connections-modifier, AV valve, Straddling, Mitral valve	06.02.09
Discordant AV connections-modifier, AV valve, Straddling, Right sided AV Valve	06.00.05
Discordant AV connections-modifier, AV valve, Straddling, Tricuspid valve	06.01.09
Discordant AV connections-modifier, Ebstein's anomaly ("Left-sided" Ebstein's anomaly) (Ebstein's malformation of 'left-sided' tricuspid valve in discordant atrioventricular connections)	06.01.75

Abbreviations: AV: atrioventricular; IVS: intact ventricular septum; LVOTO: left ventricular outflow tract obstruction; VSD: ventricular septal defect

The terminology for the first two anatomical repair options listed above has been somewhat confusing, in that they have both been referred to as double-switch procedures. The Rastelli operation, which is a part of one operation that has been erroneously called a double switch, is never referred to as a switch procedure when used in isolation in hearts with concordant atrioventricular connections. The Rastelli procedure combined with an atrial switch, therefore, should not be called a double switch. In the case of the atrial switch combined with the arterial switch, the anatomic substrate and physiologic considerations are much different. In essence, there will be minimal if any obstruction to pulmonary flow, and therefore an extracardiac ventriculo-pulmonary arterial connection is not required. Furthermore, the procedure may be performed without a co-existent ventricular septal defect. When present, simple closure of the ventricular septal defect is necessary, and the use of an intraventricular baffle is not needed. Furthermore, in almost all cases, no ventriculotomy is necessary for the closure of the ventricular septal defect when an atrial switch is combined with an arterial switch. Preparation of the morphologically left ventricle, in contrast, enabling it to function adequately as the systemic ventricle, may be an important additional issue.[43] The Senning and Mustard operations are commonly referred to as atrial switch operations. Hence, we recommend that the term "double-switch" be reserved solely for patients undergoing an atrial switch combined with an arterial switch operation. An appropriate shorthand term for the Senning or Mustard procedure combined with baffling the left ventricle to the aorta and placing a conduit from the right ventricle to the pulmonary arteries would be "Atrial switch and Rastelli". This approach is not merely semantics, as the patients submitted to surgery by these procedures are divergent groups, and comparisons of those undergoing a double switch will be meaningful only when those with associated ventricular septal defects and obstruction of the pulmonary outflow tract have been placed in their own separate category.

Nomenclature crossmap for diagnoses and therapeutic options related to discordant atrioventricular connections

In Table 1, we show the terminology for diagnoses related to discordant atrioventricular connections from the version of the International Code derived from the lists of the Society of Thoracic Surgeons and the European Association of Cardio-Thoracic Surgery. In Table 2, we show the equivalent and crossmapped terms from the version derived from the list of the Association for European Pediatric Cardiology.

Table 2. The Association for European Pediatric Cardiology derived version of the International Pediatric and Congenital Cardiac Code for the Diagnoses related to Discordant Atrioventricular Connections.

Usual atrial arrangement (atrial situs solitus)	01.03.00
Mirror image atrial arrangement (atrial situs inversus)	01.03.01
Discordant AV connections	01.04.01
Concordant VA connections	01.05.00
Congenitally corrected transposition of great arteries (discordant AV & VA connections)	01.01.03
Double outlet right ventricle	01.01.04
Criss-cross heart (twisted AV connections)	02.03.03
Aortic orifice anterior with respect to pulmonary orifice	02.06.03
Aortic orifice anterior right with respect to pulmonary orifice	02.06.02
Aortic orifice anterior left with respect to pulmonary orifice	02.06.04
Superior-inferior ventricular ("upstairs-downstairs") relationships	02.04.00
Left ventricular outflow tract obstruction	07.09.01
Left ventricular outflow tract obstruction, – not present	07.09.01 + Q1.90.81
Intact ventricular septum	07.21.00
VSD	07.10.00
Overriding AV valve	06.00.01
Overriding right AV valve	06.00.02
Overriding tricuspid valve	06.01.05
Overriding left AV valve	06.00.03
Overriding mitral valve	06.02.05
Straddling AV valve	06.00.04
Straddling right AV valve	06.00.05
Straddling tricuspid valve	06.01.09
Straddling left AV valve	06.00.06
Straddling mitral valve	06.02.09
Ebstein's malformation of 'left-sided' tricuspid valve in discordant AV connections	06.01.75

Abbreviations: AV: atrioventricular; VA: ventriculo-arterial; VSD: ventricular septal defect

Table 3 details the nomenclature for procedures related to discordant atrioventricular connections from the version of the International Code derived from the nomenclature of the International Congenital Heart Surgery Nomenclature and Database Project, while Table 4 shows the equivalent and crossmapped terms using the version of the International Code derived from the nomenclature of the European Pediatric Cardiac Code.

Summary

During the process of creation of a bidirectional crossmap between the system emerging, on the one hand, from the initiative sponsored by the Congenital Heart Committees of the European Association for Cardio-Thoracic Surgery and the Society of Thoracic Surgeons,[49] and on the other hand, from that formulated by the Coding Committee of the European Association for Pediatric Cardiology,[50,51] the Nomenclature Working Group has successfully created the International Paediatric and Congenital Cardiac Code. As would be expected, during the process of crossmapping it became clear that, for most lesions, the European Pediatric Cardiac Code was more complete in its description of the diagnoses, while the International Congenital Heart Surgery Nomenclature and Database Project was more complete in its description of the procedures. This process[2–7] of crossmapping exemplifies the efforts of the Nomenclature Working Group to create a comprehensive and all-inclusive international system for the naming of paediatric and congenital cardiac disease, the International Pediatric and Congenital Cardiac Code. Although names and classification for paediatric and congenital cardiac disease will continue to evolve over time, we are now closer than ever to reaching uniform international agreement and standardization. The International Paediatric and Congenital Cardiac Code can be downloaded from the Internet, free of charge, at www.ipccc.net.

Table 3. The European Association for Cardiothoracic Surgery – Society of Thoracic Surgeons derived version of the International Pediatric and Congenital Cardiac Code for Procedures related to Discordant Atrioventricular Connections.

Congenitally corrected TGA repair	12.27.46
Congenitally corrected TGA repair, Anatomic repair, Atrial switch and Arterial switch (Double switch)	12.29.25
Congenitally corrected TGA repair, Anatomic repair, Atrial switch and Intraventricular tunnel repair, Reversed tunnel repair of Patrick and McGoon	12.27.47
Congenitally corrected TGA repair, Anatomic repair, Atrial switch and Nikaidoh procedure (Aortic root translocation over left ventricle)	12.27.48, 12.29.06
Congenitally corrected TGA repair, Anatomic repair, Atrial switch and Rastelli (Left ventricle to aorta intraventricular tunnel repair and right ventricle to pulmonary artery conduit)	12.29.26
Congenitally corrected TGA repair, Classic repair, VSD closure	12.27.46, 12.08.01
Congenitally corrected TGA repair, Classic repair, VSD closure and LV to PA conduit	12.27.46, 12.08.01, 12.36.02
Congenitally corrected TGA repair, Fontan	12.30.01
Congenitally corrected TGA repair, One-and-one half ventricular repair	12.06.49
Congenitally corrected TGA repair, One-and-one half ventricular repair, VSD closure + Superior cavopulmonary anastomosis (Glenn anastomosis)	12.06.49, 12.08.01
Congenitally corrected TGA repair, Other	12.27.47
Congenitally corrected TGA repair, Staged procedure ("2 stage anatomic repair")	12.27.46, 12.29.24
Congenitally corrected TGA repair, Staged procedure ("2 stage anatomic repair"), Atrial switch and Arterial switch (Double switch) Status post preceding PA banding ("2 stage switch")	12.27.46, 12.29.24, 12.29.25, 12.14.02 + Q5.81.17
Congenitally corrected TGA repair, Staged procedure ("2 stage anatomic repair"), Atrial switch and Rastelli Status post preceding PA banding	12.27.46, 12.29.24, 12.29.26, 12.14.02 + Q5.81.17
Congenitally corrected TGA repair-modifier, With atrioventricular valve repair	12.90.01
Congenitally corrected TGA repair-modifier, With atrioventricular valve replacement	12.90.02
Atrial switch procedure	12.29.06
Atrial switch procedure, Mustard procedure	12.29.02
Atrial switch procedure, Mustard procedure, With PA Debanding	12.29.02, 12.14.03
Atrial switch procedure, Mustard procedure, With subpulmonic obstruction repair (LVOTO repair)	12.29.02, 12.07.13
Atrial switch procedure, Mustard procedure, With subpulmonic obstruction repair (LVOTO repair) and VSD closure	12.29.02, 12.07.13, 12.08.01
Atrial switch procedure, Mustard procedure, With VSD closure	12.29.02, 12.08.01
Atrial switch procedure, Non-Mustard – non-Senning	12.29.19
Atrial switch procedure, Senning procedure	12.29.01
Atrial switch procedure, Senning procedure, With PA Debanding	12.29.01, 12.14.03
Atrial switch procedure, Senning procedure, With subpulmonic obstruction repair (LVOTO repair)	12.29.01, 12.07.13
Atrial switch procedure, Senning procedure, With subpulmonic obstruction repair (LVOTO repair) and VSD closure	12.29.01, 12.07.13, 12.08.01
Atrial switch procedure, Senning procedure, With VSD closure	12.29.01, 12.08.01
Atrial switch procedure, Senning procedure-modifier, Classical Senning with no patch	12.29.04
Atrial switch procedure, Senning procedure-modifier, Senning with left atrial appendage involved (Inverted left atrial appendage)	12.29.18
Atrial switch procedure, Senning procedure-modifier, Senning with patch	12.29.07

Abbreviations: LV: left ventricle; PA: pulmonary arterial; TGA: transposition of the great arteries; VSD: ventricular septal defect

Table 4. The Association for European Paediatric Cardiology derived version of the International Paediatric and Congenital Cardiac Code for the Procedures related to Discordant Atrioventricular Connections.

Congenitally corrected transposition of great arteries repair	12.27.46
Arterial switch & atrial inversion procedures ("double switch")	12.29.25
Atrial inversion and Rastelli procedures	12.29.26
Aortic root posterior translocation to left ventricle + biventricular outflow tract reconstruction (Nikaidoh)	12.27.48
Atrial inversion procedure & intraventricular reversed tunnel repair (Patrick-McGoon)	12.29.27
Congenitally corrected transposition of great arteries repair (DESCRIBE)	12.27.47
Atrial inversion procedure	12.29.06
Senning procedure (atrial inversion)	12.29.01
Senning procedure (atrial inversion): without patch (classic)	12.29.04
Senning procedure (atrial inversion): with patch	12.29.07
Senning procedure (atrial inversion): using inverted left atrial appendage	12.29.18
Mustard procedure (atrial inversion)	12.29.02
Atrial inversion procedure (non-Mustard or Senning)	12.29.19
Arterial switch procedure: staged	12.29.24
1.5 ventricle repair	12.06.49
Fontan type procedure	12.30.01
Left ventricle to pulmonary artery conduit construction	12.36.02
VSD closure	12.08.01
Left ventricular outflow tract obstruction relief	12.07.13
AV valvar repair	12.90.01
AV valvar replacement	12.90.02
Pulmonary trunk band (PA band), – preceding	12.14.02 + Q5.81.17
Pulmonary trunk band removal (de-band)	12.14.03

Abbreviations: As for Table 3

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