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Classification of the functionally univentricular heart: unity from mapped codes

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THE NOMENCLATURE AND CLASSIFICATION OF patients with a functionally univentricular heart has been debated for decades. We review here the approach taken for dealing with this group of patients by the International Working Group for Mapping and Coding of Nomenclatures for Paediatric and Congenital Cardiac Disease. We discuss the approach of this Nomenclature Working Group in the context of other historical and contemporary ideas about this topic.

Historical concepts

The debate about the proper nomenclature for the functionally univentricular heart goes back several decades. The approach taken at Boston Children's Hospital from the 1970s is summarized by the following passage, authored by Donald C. Fyler, in the textbook "Nadas' Pediatric Cardiology":[1]

> "At Boston Children's Hospital, a single ventricle is defined as the presence of two atrioventricular valves with one ventricular chamber or a large dominant ventricle associated with a diminutive opposing ventricle.[2–4] The term double-inlet ventricle is also used to describe this group of anomalies. While the concept of a univentricular heart[5–8] fits the physiologic idea of a common mixing chamber and has pathologic merits as well, lumping patients with mitral atresia, tricuspid atresia, and others into one category adds confusion rather than contributing to classification. The easy distinction between a single atrioventricular valve and two atrioventricular valves has been a reproducible basis for clinical impressions extending over many years. To change nomenclature would require significant benefits that are not apparent at the present time."

The concept of the "univentricular heart" as discussed above by Fyler is essentially the one evolved by a group of European morphologists and clinicians, albeit with strong support from Freedom in Toronto.[5–7] The evolution of their system, however,

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depended on emphasizing that, in the hearts under discussion, it was the atrioventricular connection, rather than the heart itself, which was truly univentricular. Thus, in one of the cited works,[8] the situation was summarized as follows:

> "In most hearts with double-inlet connection it is not the ventricles that are univentricular; it is the AV connection. The concept of a univentricular AV connection, then, appropriately groups hearts with double-inlet along with those having absence of one AV connection. It distinguishes this entire group from those other hearts with biventricular AV connections (each atrium connected to its own ventricle). The term "univentricular AV connection" is thus a collective one for all those hearts in which the atria connect to only one ventricle."

With ongoing experience, it has now become clear that many of these hearts with biventricular atrioventricular connections can also lead to a situation which, in terms of physiology, is functionally univentricular. Thus, in the initial review of this supplement, Jacobs and Anderson[9] explain how the term "functionally univentricular heart" allows for the grouping together of hearts in which "one chamber was incapable independently of supporting either the pulmonary or the systemic circulation". This approach is based upon the concept of "*appreciating that the entire ventricular mass was functionally univentricular whenever one or other ventricle was incapable, for whatever reason, of supporting either the systemic or the pulmonary circulation*".[9] The endpoint of this evolutionary journey, therefore, is an approach that permits description of the patients possessing functionally univentricular hearts based upon a clear understanding of the cardiac phenotype, and a detailed description of this cardiac phenotype.

Current approaches for creation of databases: The International Paediatric and Congenital Cardiac Code

There can be little argument that the best methodology to describe any given heart is a detailed description of the cardiac phenotype, as advocated elsewhere in this supplement.[9] In the framework of the multi-institutional database, nonetheless, pure description of the cardiac phenotype is best augmented by a system of classification that allows grouping of common lesions for various analyses and research studies.

In 1998, the Society of Thoracic Surgeons, together with the European Association for Cardio-Thoracic Surgery, established the International Congenital Heart Surgery Nomenclature and Database Project, seeking to create a common nomenclature and database for congenital cardiac malformations. Such a proposed common nomenclature, along with a common core representing the minimal dataset, were adopted by these two societies and published in 2000.[10] Also in 2000, however, the Association for European Paediatric Cardiology published their suggested system for nomenclature, specifically the European Paediatric Cardiac Code.[11] In the 5 years since this time, the Nomenclature Working Group,[12–17] established during the World Congress held in Toronto in 2001, has worked to unify these two systems. This has been accomplished by bidirectional crossmapping of the two systems to a common numerical backbone,[10,11] with additional mapping to incorporate the lists developed for the Canadian Congenital Heart Codes, as yet unpublished, the codes developed in Boston by Donald C. Fyler, and also as yet unpublished, and the 9th and 10th revisions of the International Classification of Diseases, of the World Health Organization. The product of this crossmapping is the International Paediatric and Congenital Cardiac Code, which consists of two versions: the surgical version based upon the International Congenital Heart Surgery Nomenclature and Database Project,[10] and the European cardiology version based upon the European Paediatric Cardiac Code.[11] Both versions, in fact, have had considerable input from paediatric cardiologists and cardiac surgeons. The crossmapping was presented at the Second International Summit on Nomenclature for Pediatric and Congenital Heart Disease at The Fourth World Congress of Pediatric Cardiology and Cardiac Surgery, held in Buenos Aires, Argentina, over the period September 18 through 22, 2005, and is available for free download from the Internet at www.ipccc.net.[18]

In this review, we have extracted from the International Paediatric and Congenital Cardiac Code the section developed for patients with functionally univentricular hearts. In order to appreciate this nomenclature, it is necessary first understand the previously published principles and rules relating to four areas of crossmapping:[16]

- Generic terms in the lists, that is terms ending in *NOS* in the surgical lists or (*unspecified*) in the European lists.
- Nonspecific terminology meant to allow further description in the nomenclature lists, that is terms ending in *Other* in the surgical lists or (*DESCRIBE*) in the European lists.
- The meaning of the words *right* and *left*.
- Structural differences between the two systems of nomenclature.

Optimal performance from systems of nomenclature can be expected in an environment where the database, or system for entry of data, has certain standard regulations and requirements. The person entering the data, the coder, must be forced to

choose from the choices in the list of nomenclatures, and not be allowed to type free text directly into the fields for "Diagnoses" and "Procedures". A separate "Comments" field will then allow further free text to add additional description to any individual diagnosis or procedure that has been chosen. The crossmapping, and the systems themselves, will work effectively in environments that follow this basic rule or principle.

This fundamental principle also leads to logical solutions for the first two issues highlighted above.

All terms in the nomenclature lists theoretically end in *NOS* or (*unspecified*), in that one can always create further subdivisions for virtually any diagnosis or procedure. As stated above, therefore, the generic term on its own is self explanatory, without the need for other clarifying nomenclature, such as *NOS* or (*unspecified*) being affixed. These suffixes are consequently not necessary.

The terms ending in *Other* in the surgical lists are problematic for several reasons. The appendage *Other* could confer different meanings to a term depending on the list in which it is included, and any entry containing the appended term *Other* may change meaning over time, as additional terms are added to the parent list from which the term is derived. The purpose and original intent of these appended terms in the surgical lists was to allow for the further description of related terms or choices not appearing in the list, similar to the use of the suffix (*DESCRIBE*) in the European lists. The initial solution proposed for the discrepancy between terms ending in *Other* in the surgical lists, and (*DESCRIBE*) in the European lists, was to convert the terms ending in *Other* in the surgical lists to (*DESCRIBE*), as this would circumvent the above shortcomings and implications inherent in the word *Other*. It is apparent, however, that when the database environment follows the rule discussed above, namely, that no free text is permitted in the fields for "Diagnoses" or "Procedures", then there is no longer a requirement to specify that a family of terms can have further items added. A separate "Comments" field exists to allow further description of any chosen item. Thus, theoretically, all terms in the lists are suffixed with (*DESCRIBE*), and the coder has the option to add further detail to any selected term. As a consequence, generic family terms ending in (*DESCRIBE*) or *Other* become redundant.

When discussing cardiac chambers, such as atriums and ventricles, and spatial relationships, the words *left* and *right* can be confusing. Rules were created, therefore, to provide consistency and accuracy of descriptive terms of anatomical phenotypes. For cardiac chambers, unless otherwise stated, *left* refers to morphologically left, and *right* refers to morphologically right. Thus, left ventricle means the morphologically left ventricle, left atrium refers to the morphologically left atrium, and right atrial appendage refers to the morphologically right atrial appendage, and so on. With regard to the third item, when discussing cardiac chambers, the words *left* and *right* do not imply sidedness or position. If the position or sidedness of a cardiac chamber is to be described, it is necessary to use terms such as *left-sided ventricle*. The term "left ventricle", therefore, merely means the morphologically left ventricle, and does not mean or imply left-sidedness or right-sidedness. Similarly, it does not imply connections to the right or left atrium, or the pulmonary or systemic circulations. In contrast, when describing the superior caval vein, and using the prefix *left* or *right*, it is the spatial position that is being alluded to, rather than any other connection or phenotypic variation that may exist.

The fourth issue evolves from the fact that the structure of the two systems for nomenclature differs fundamentally. This difference is most apparent when comparing the two Long Lists. The nomenclature developed by the International Congenital Heart Surgery Nomenclature and Database Project Committee uses a tree for its hierarchical structure, with an incrementally more complex diagnostic or procedural combination of terms. Each combination is considered a single diagnostic unit, which theoretically would have its own numerical code, had the Committee chosen to create a system of numbers. In contrast, the European Paediatric Cardiac Code is largely constructed in an "atomic" way, so that a complex diagnosis would have separate numerical codes for each element. This means that a map between the two systems leads to a series of codes in the European Code being equivalent to one "unit" of diagnosis in the Surgical Code. The combination from the surgical nomenclature "TGA, VSD – LVOTO" is equivalent to three entries in the European Paediatric Cardiac Code, specifically "Discordant VA connections (01.05.01)", "VSD (07.10.00)", and "Left ventricular outflow tract obstruction (07.09.01)". In the mapping of the Short Lists, this has been addressed by "boxing" together groups of terms from the European Paediatric Cardiac Code, and listing them at the end of the crossmap of the European Paediatric Cardiac Code to the International Congenital Heart Surgery Nomenclature and Database Project as an Appendix, whilst integrating them into the structure of the reverse crossmap. Exceptions to this configuration are a few common combinations of lesions that are so routinely associated with each other that they have been grouped as one discrete diagnosis or procedure in both systems. Examples[16] are "Pulmonary atresia+VSD (including Fallot type) (01.01.06)", or "Arterial and atrial switch procedures (double switch) (12.29.25)."

The Long List established by the surgeons, therefore, is structured on the basis of "molecules" of terms

commonly found together, whilst the long list of the European Paediatric Cardiac Code takes an atomic approach, and lists each diagnostic element separately.

The surgical Long List thus created, however, is not intended to be all inclusive of every possible molecule. Instead, the intention is to list the more common diagnostic groupings as diagnostic molecules. This surgical Long List, nonetheless, does provide many of the diagnostic atoms needed to code rarer lesions using the atomic approach present in the European Pediatric Cardiac Code. The difference in the two approaches is not problematic. Indeed, it is to be expected, and predicted, by the purpose of each list. The Surgical Long List is trying to allow for common diagnostic and procedural groupings to be grouped together for multi-institutional analysis of databases. The European code was established to be used as a database system and to permit the description of combinations of lesions that are unusual or may never have been previously encountered. Both systems allow for patients with rarer lesions to be coded with the "atomic" approach. In the situation where a patient may be diagnosed with an element that does not have a code, either system will allow for further description to be added in the section provided for "Diagnostic Comments".

The newly developed International Paediatric and Congenital Cardiac Code now provides a common numerical backbone of crossmapped terms that can be accessed from multiple different diagnostic listings. As already discussed, in this review we will highlight the listings developed for patients possessing functionally univentricular hearts.

The Version of the International Paediatric and Congenital Cardiac Code for the Functionally Univentricular Heart Derived from the Nomenclature of the European Association for Cardio-Thoracic Surgery and the Society of Thoracic Surgeons

The system uses the term "single ventricle" as synonymous for the functionally univentricular heart. When first published, it was stated that:

> "Personal communication with Professor Robert Anderson at the 13th Annual Meeting of The European Association for Cardio-Thoracic Surgery, Glasgow, Scotland, September 5–8, 1999 reveals that Professor Anderson prefers the term "functionally single ventricle" rather than the term "single ventricle" because these hearts generally have a functional single ventricle in addition to a diminutive or hypoplastic ventricle. We agree that the hearts that we classify as single ventricle in reality have a single well-developed ventricle and may also have an additional incomplete, rudimentary, or hypoplastic ventricle. Thus, our concept of single ventricle is consistent with Professor Anderson's concept of functionally single ventricle. Our reluctance to use the term functionally single ventricle in this database scheme stems from the popular use of the term single ventricle in the surgical literature. As these initiatives progress, more debate by surgeons, anatomists, and pediatric cardiologists may result in nomenclature changes that will keep these initiatives as works in progress with the eventual goal of establishing a uniform nomenclature system across geographic boundaries and specialty preferences."

Patients classified in this section of the nomenclature, therefore, include all those who would be coded using the Short List for "Single Ventricle", specifically:

- Single ventricle 01.01.22
- Single ventricle, DILV 01.04.04
- Single ventricle, DIRV 01.04.03
- Single ventricle, Heterotaxia syndrome 01.01.22 + 03.01.02
- Single ventricle, Mitral atresia 01.01.22 + 06.02.01
- Single ventricle, Tricuspid atresia 01.01.22 + 06.01.01
- Single ventricle, Unbalanced AV canal 01.01.22 + 06.07.26

This Short List expands to the Long List as shown in Table 1. The Long List itself then has a separate section allowing for the detailed coding of patients with either hypoplastic left heart syndrome (hypoplasia of the left heart), or pulmonary atresia in the setting of an intact ventricular septum. Most of the patients in the first of these groupings, and many of those in the second, are likely to be repaired surgically in a functionally univentricular fashion. These two subsets with hypoplasia of the left or right hearts, nonetheless, also possess their own detailed breakdown within the nomenclature. The person responsible for analyzing the data, therefore, always has the opportunity to group patients coded in this part of the system with those coded explicitly as possessing functionally univentricular hearts for any given analysis. As explained previously:

> *The consensus of the Society of Thoracic Surgeons Congenital Heart Surgery Database Committee and representatives from the European Association for Cardio-Thoracic Surgery was that the nomenclature proposal for single ventricle hearts would encompass hearts with double inlet atrioventricular connection (both double inlet left ventricle (DILV) and double inlet right ventricle (DIRV)), hearts with absence of one atrioventricular connection (mitral atresia and tricuspid atresia), hearts with a common atrioventricular valve and only one completely well-developed ventricle (unbalanced*

common atrioventricular canal defect), hearts with only one fully well-developed ventricle and heterotaxia syndrome (single ventricle heterotaxia syndrome), and finally other rare forms of univentricular hearts that do not fit in one of the specified major categories. Despite the recognition that hypoplastic left heart syndrome is a common form of univentricular heart, with a single or dominant ventricle of right ventricular morphology, the current nomenclature and database proposal includes an entirely separate section for consideration of hypoplastic left heart syndrome. Also, it is recognized that a considerable variety of other structural cardiac malformations, such as pulmonary atresia with intact ventricular septum, biventricular hearts with straddling atrioventricular valves, and some complex forms of double outlet right ventricle (DORV), may at times be best managed in a fashion similar to that which is used to treat univentricular hearts. Nomenclature for description of those entities, however, is not included in this section."[19]

The original Short List and Long List developed by the International Congenital Heart Surgery Nomenclature and Database Project Committee[10] has now been utilized in multiple multi-institutional research studies. For example:

- The databases of the members of the European Association for Cardio-Thoracic Surgery and the Society of Thoracic Surgeons which currently track data relative to outcomes from over 50000 surgical procedures carried out in Europe and North America.[20–25]
- A multi-institutional study of functionally single ventricle via the Pediatric Heart Network.
- The Centers for Disease Control and Prevention birth surveillance research study in which The Metropolitan Atlanta Congenital Defects Program has reclassified more than 11,000 patients according to the surgical system.
- A National Institute of Health grant looking at the relationship of air pollution to the development of congenital cardiac malformations in the fetus (R01ES012967).
- The trial funded by the National Institute of Health, and conduced by the Pediatric Heart Network, comparing construction of a conduit placed from the right ventricular to the pulmonary arteries as opposed to the modified Blalock–Taussig shunt in infants undergoing staged reconstruction with functionally univentricular hearts and hypoplastic left heart syndrome.
- Various software houses have incorporated the Long and Short Lists into their systems to facilitate the above programs.

The original surgical Long List for "single ventricle" has now been modified and improved as a result of the crossmapping and creation of the International Paediatric and Congenital Cardiac Code (Table 1). These modifications and improvements were all done in a framework that will allow for retention of the structure of the initial database and the research it has spawned thus far.

It should be noted, nonetheless, that some patients with atrioventricular valvar atresia can have biventricular atrioventricular connections, yet still be functionally univentricular. It would be inappropriate to code these within the items offered in Table 1. Patients recognized with such lesions must be described and coded atomically, using any of multiple available diagnostic codes, including the following terms taken from the surgical Long List:

AV connection = Absent left sided AV connection
AV connection = Absent left sided AV connection – univentricular
AV connection = Absent left sided AV connection with straddling valve – uniatrial biventricular
AV connection = Absent left sided AV connection, Right sided atrium to both ventricles
AV connection = Absent left sided AV connection, Right sided atrium to LV
AV connection = Absent left sided AV connection, Right sided atrium to RV
AV connection = Absent left sided AV connection, Right sided atrium to ventricle of indeterminate morphology
AV connection = Absent right sided AV connection
AV connection = Absent right sided AV connection – univentricular
AV connection = Absent right sided AV connection with straddling valve – uniatrial biventricular
AV connection = Absent right sided AV connection, Left sided atrium to both ventricles
AV connection = Absent right sided AV connection, Left sided atrium to LV
AV connection = Absent right sided AV connection, Left sided atrium to RV
AV connection = Absent right sided AV connection, Left sided atrium to ventricle of indeterminate morphology

AV valve, Imperforate
AV valve, Imperforate, Left sided AV valve
AV valve, Imperforate, Mitral valve
AV valve, Imperforate, Right sided AV valve
AV valve, Imperforate, Tricuspid valve

AV valve overriding
AV valve overriding, Left sided AV valve
AV valve overriding, Mitral valve
AV valve overriding, Right sided AV valve
AV valve overriding, Tricuspid valve

AV valve overriding-modifier for degree of override
AV valve overriding-modifier for degree of override, Override of AV valve <50%
AV valve overriding-modifier for degree of override, Override of AV valve >90%
AV valve overriding-modifier for degree of override, Override of AV valve 50–90%

AV valve straddling
AV valve straddling, Left sided AV valve
AV valve straddling, Mitral valve
AV valve straddling, Right sided AV valve
AV valve straddling, Tricuspid valve.

The Version of the International Paediatric and Congenital Cardiac Code for the Functionally Univentricular Heart Derived from the European Paediatric Cardiac Code of the Association for European Paediatric Cardiology

The version of the International Paediatric and Congenital Cardiac Code derived from the European Paediatric Cardiac Code is based on an atomic approach that allows explicit description of the cardiac phenotype. In line with the previous discussion, the structure of the European Paediatric Cardiac Code necessitates that, for the most part, combinations of individual codes are used to describe the possible variants of the functionally univentricular heart, with appropriate qualifiers. In contrast to the version prepared for the European Association for Cardio-Thoracic Surgery and the Society of Thoracic Surgeons, the version used by the European Association for Paediatric Cardiology does not subcategorize the various entities described as having a functionally univentricular heart under this specific heading. Rather, this description can be used as an additional descriptor if so wished, using the following terms:

Functionally univentricular heart	01.01.22
Ventricular imbalance	07.08.40
Ventricular imbalance: dominant left ventricle + hypoplastic right ventricle	07.08.41
Ventricular imbalance: dominant right ventricle + hypoplastic left ventricle	07.08.42

The specific lesions are then preferably described independently and in their own right:

Double inlet ventricle	01.01.14
Tricuspid atresia	06.01.01
Mitral atresia	06.02.01
Pulmonary atresia + intact ventricular septum	01.01.07
Hypoplastic left heart syndrome	01.01.09
Solitary ventricle of indeterminate morphology	02.03.05

Of importance is that the version derived from the European Paediatric Cardiac Code uses the sequential segmental approach[26–28] to describe these cardiac malformations by building up the structure of the lesion: atrial arrangement (situs), atrioventricular connection(s), and ventriculo-arterial connection(s), followed by descriptors of various additional anomalies, such as a ventricular septal defect or obstruction within the outflow tracts. Thus, the terms detailed in Table 2 need to be used as a cumulative combination of terms to equate with the lesions listed in Table 1. Both Table 1 and Table 2 are clearly incomplete, as they do not exhaustively describe the many variations of morphology which may be found in hearts producing functionally univentricular physiology. For instance, anomalies of the pulmonary venous connections, and lesions of the aortic arches, are not listed. They need to be coded separately in both the new versions of the International Paediatric and Congenital Cardiac Code.

The revised Short and Long Lists of the European Paediatric Cardiac Code[29] have been downloaded from the internet more than 600 times.[30] The lists have, and are, being used in multiple multi-institutional studies throughout Europe:

- In the United Kingdom, the Central Cardiac Audit Database uses the Short List as the basis for its national, comprehensive, validated and benchmark driven audit of all paediatric surgical and transcatheter procedures undertaken since 2000, with over 28,000 procedures entered to date.[31]
- Internal quality control for all centres in Germany, with either the Short or Long lists used depending upon the centre.
- In Germany, the "Nationale Register für angeborene Herzfehler" in Berlin uses the Short List for coding all patients with congenital heart disease in Germany – an epidemiological study with over 15,000 patients registered to date.[32]
- In Germany, the "Kompetenznetz angeborene Herzfehler" uses the Short List for a nation-wide scientific network supported by the German government for various specific studies, such as right ventricular function, pulmonary hypertension, tetralogy of Fallot, interatrial communication and so on.[33]
- In the Netherlands, the national registry of congenital heart disease CONCOR (CONgenital CORvitia) has over 6000 patients registered using the Short List.[34]
- The Swiss paediatric cardiology society uses the European Paediatric Cardiac Code Short List for quality control between centres. This is independent and nongovernmental.
- Various software houses have incorporated the Long and Short Lists into their systems to facilitate the above programs.

Summary

Since 2001, the International Working Group for Mapping and Coding of Nomenclatures for Paediatric and Congenital Cardiac Disease has been working to unify the two distinct systems developed to promote international nomenclature by mapping them to a common numerical backbone. This has resulted in the versions of the International Paediatric and Congenital Cardiac Code derived, on the one hand, from the system initially developed by the European Association for Cardio-Thoracic Surgery and the Society of Thoracic Surgeons, and on the other hand by the Association for European Paediatric Cardiology. This article establishes that this process of unification has been feasible even for such a complex and controversial family of lesions subjectively united by the concept of a functionally univentricular heart. Comparison of the two tables demonstrates that the fundamentally different approaches to nomenclature, one atomic and the other molecular, originally taken by the European Association for Cardio-Thoracic Surgery and the Society of Thoracic Surgeons and the Association for European Paediatric Cardiology in 2000 can be mapped to generate the same numerical codes which are lesion-specific. Either version of the International Paediatric and Congenital Cardiac Code will now allow the coding and description of patients possessing functionally univentricular hearts. With the addition of atomic modifiers and qualifiers, either system will allow for the additional detailed description of the cardiac phenotype, as advocated by Jacobs and Anderson.[9] It must be remembered, for example, that patients with biventricular atrioventricular connections and imperforate atrioventricular valves do rarely present with atrioventricular valvar atresia. These rarer hearts will be coded in either list with specific anatomic terms to describe the cardiac phenotype. Only by recognizing such rarities will we identify eventually the range of anatomic risk factors that contribute to the success or failure of surgical procedures. By sharing a common numerical backbone, nonetheless, these two versions of the International Paediatric and Congenital Cardiac Code make it possible for practitioners using either system to communicate with each other. The aim is to facilitate the detailed description of the cardiac phenotype, while at the same time facilitating meaningful multi-institutional research.

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Table 1. The European Association for Cardio-Thoracic Surgery – Society of Thoracic Surgeons version of the International Paediatric and Congenital Cardiac Code for the Functionally Univentricular Heart.

Single ventricle	01.01.22
Single ventricle, DILV	01.04.04
Single ventricle, DILV, {SDD}	01.04.04, 01.03.00, 02.03.01, 02.06.02, 01.05.01
Single ventricle, DILV, {SDD}, Subaortic RV outlet chamber with VSD (Bulboventricular foramen)	01.04.04, 01.03.00, 02.03.01, 02.06.02, 01.05.01, 07.02.00, 07.10.00
Single ventricle, DILV, {SDD}, Subaortic RV outlet chamber with VSD (Bulboventricular foramen), Nonrestrictive bulboventricular foramen	01.04.04, 01.03.00, 02.03.01, 02.06.02, 01.05.01, 07.02.00, 07.14.04
Single ventricle, DILV, {SDD}, Subaortic RV outlet chamber with VSD (Bulboventricular foramen), Nonrestrictive bulboventricular foramen and no pulmonary atresia and no pulmonary stenosis	01.04.04, 01.03.00, 02.03.01, 02.06.02, 01.05.01, 07.02.00, 07.14.04, 09.05.94
Single ventricle, DILV, {SDD}, Subaortic RV outlet chamber with VSD (Bulboventricular foramen), Nonrestrictive bulboventricular foramen and pulmonary atresia	01.04.04, 01.03.00, 02.03.01, 02.06.02, 01.05.01, 07.02.00, 07.14.04, 09.05.11
Single ventricle, DILV, {SDD}, Subaortic RV outlet chamber with VSD (Bulboventricular foramen), Nonrestrictive bulboventricular foramen and pulmonary stenosis	01.04.04, 01.03.00, 02.03.01, 02.06.02, 01.05.01, 07.02.00, 07.14.04, 09.05.92
Single ventricle, DILV, {SDD}, Subaortic RV outlet chamber with VSD (Bulboventricular foramen), Restrictive bulboventricular foramen	01.04.04, 01.03.00, 02.03.01, 02.06.02, 01.05.01, 07.02.00, 07.14.01
Single ventricle, DILV, {SDD}, Subaortic RV outlet chamber with VSD (Bulboventricular foramen), Restrictive bulboventricular foramen and no pulmonary atresia and no pulmonary stenosis	01.04.04, 01.03.00, 02.03.01, 02.06.02, 01.05.01, 07.02.00, 07.14.01, 09.05.94
Single ventricle, DILV, {SDD}, Subaortic RV outlet chamber with VSD (Bulboventricular foramen), Restrictive bulboventricular foramen and pulmonary atresia	01.04.04, 01.03.00, 02.03.01, 02.06.02, 01.05.01, 07.02.00, 07.14.01, 09.05.11
Single ventricle, DILV, {SDD}, Subaortic RV outlet chamber with VSD (Bulboventricular foramen), Restrictive bulboventricular foramen and pulmonary stenosis	01.04.04, 01.03.00, 02.03.01, 02.06.02, 01.05.01, 07.02.00, 07.14.01, 09.05.92
Single ventricle, DILV, {SDN} ({SDS}) (Subpulmonary RV outlet chamber)	01.04.04, 01.03.00, 02.03.01, 01.05.00, 07.02.00
Single ventricle, DILV, {SDN} ({SDS}) (Subpulmonary RV outlet chamber), (Holmes heart)	01.04.04, 01.03.00, 02.03.01, 01.05.00, 07.02.00
Single ventricle, DILV, {SLL}	01.04.04, 01.03.00, 02.03.02, 02.06.04, 01.05.01
Single ventricle, DILV, {SLL}, Subaortic RV outlet chamber with VSD (Bulboventricular foramen)	01.04.04, 01.03.00, 02.03.02, 02.06.04, 01.05.01, 07.02.00, 07.10.00
Single ventricle, DILV, {SLL}, Subaortic RV outlet chamber with VSD (Bulboventricular foramen), Nonrestrictive bulboventricular foramen	01.04.04, 01.03.00, 02.03.02, 02.06.04, 01.05.01, 07.02.00, 07.14.04
Single ventricle, DILV, {SLL}, Subaortic RV outlet chamber with VSD (Bulboventricular foramen), Nonrestrictive bulboventricular foramen and no pulmonary atresia and no pulmonary stenosis	01.04.04, 01.03.00, 02.03.02, 02.06.04, 01.05.01, 07.02.00, 07.14.04, 09.05.94
Single ventricle, DILV, {SLL}, Subaortic RV outlet chamber with VSD (Bulboventricular foramen), Nonrestrictive bulboventricular foramen and pulmonary atresia	01.04.04, 01.03.00, 02.03.02, 02.06.04, 01.05.01, 07.02.00, 07.14.04, 09.05.11
Single ventricle, DILV, {SLL}, Subaortic RV outlet chamber with VSD (Bulboventricular foramen), Nonrestrictive bulboventricular foramen and pulmonary stenosis	01.04.04, 01.03.00, 02.03.02, 02.06.04, 01.05.01, 07.02.00, 07.14.04, 09.05.92
Single ventricle, DILV, {SLL}, Subaortic RV outlet chamber with VSD (Bulboventricular foramen), Restrictive bulboventricular foramen Restrictive bulboventricular foramen	01.04.04, 01.03.00, 02.03.02, 02.06.04, 01.05.01, 07.02.00, 07.14.01
Single ventricle, DILV, {SLL}, Subaortic RV outlet chamber with VSD (Bulboventricular foramen), Restrictive bulboventricular foramen and no pulmonary atresia and no pulmonary stenosis	01.04.04, 01.03.00, 02.03.02, 02.06.04, 01.05.01, 07.02.00, 07.14.01, 09.05.94

(Continued)

Table 1. (Continued)

Single ventricle, DILV, {SLL}, Subaortic RV outlet chamber with VSD (Bulboventricular foramen), Restrictive bulboventricular foramen and pulmonary atresia	01.04.04, 01.03.00, 02.03.02, 02.06.04, 01.05.01, 07.02.00, 07.14.01, 09.05.11
Single ventricle, DILV, {SLL}, Subaortic RV outlet chamber with VSD (Bulboventricular foramen), Restrictive bulboventricular foramen and pulmonary stenosis	01.04.04, 01.03.00, 02.03.02, 02.06.04, 01.05.01, 07.02.00, 07.14.01, 09.05.92
Single ventricle, DILV, DOLV	01.04.04, 01.05.03
Single ventricle, DILV, DORV	01.04.04, 01.01.04
Single ventricle, DIRV	01.04.03
Single ventricle, DIRV, DOLV	01.04.03, 01.05.03
Single ventricle, DIRV, DORV	01.04.03, 01.01.04
Single ventricle, DIRV, Subaortic LV outlet chamber with VSD ("Bulboventricular foramen")	01.04.03, 01.05.00, 07.07.00, 07.10.00
Single ventricle, DIRV, Subaortic LV outlet chamber with VSD ("Bulboventricular foramen"), Nonrestrictive VSD	01.04.03, 01.05.00, 07.07.00,07.14.04
Single ventricle, DIRV, Subaortic LV outlet chamber with VSD ("Bulboventricular foramen"), Restrictive VSD	01.04.03, 01.05.00, 07.07.00, 07.14.01
Single ventricle, Double inlet ventricle (DIV) of indeterminate ventricular morphology	01.04.05
Single ventricle, Heterotaxia syndrome	01.01.22, 03.01.02
Single ventricle, Heterotaxia syndrome, DORV, CAVC (CAVSD), Asplenia (Right isomerism)	01.01.22, 03.01.02, 01.01.04, 06.06.00, 03.01.04
Single ventricle, Heterotaxia syndrome, DORV, CAVC (CAVSD), Polysplenia (Left isomerism)	01.01.22, 03.01.02, 01.01.04, 06.06.00, 03.01.05
Single ventricle, Heterotaxia syndrome, Single LV	01.01.22, 03.01.02, 02.04.08
Single ventricle, Mitral atresia	01.01.22, 06.02.01
Single ventricle, Mitral atresia, {SDN}	01.01.22, 06.02.01, 01.03.00, 02.03.01, 01.05.00
Single ventricle, Mitral atresia, {SLL} (Corrected transposition and right-sided atrioventricular valve atresia)	**01.01.22, 01.03.00, 01.01.03, 06.02.02, 02.06.04, 02.03.02
Single ventricle, Mitral atresia, {SLL} (Corrected transposition and right-sided atrioventricular valve atresia), No Pulmonary atresia or pulmonary stenosis	**01.01.22, 01.03.00, 01.01.03, 06.02.02, 02.06.04, 02.03.02, 09.05.94
Single ventricle, Mitral atresia, {SLL} (Corrected transposition andright-sided atrioventricular valve atresia), Pulmonary atresia	**01.01.22, 01.03.00, 01.01.03, 06.02.02, 02.06.04, 02.03.02, 09.05.11
Single ventricle, Mitral atresia, {SLL} (Corrected transposition and right-sided atrioventricular valve atresia), Pulmonary stenosis	**01.01.22, 01.03.00, 01.01.03, 06.02.02, 02.06.04, 02.03.02, 09.05.92
Single ventricle, Mitral atresia, DORV	01.01.22, 06.02.01, 01.01.04
Single ventricle, Mitral atresia-modifier, Dominant left ventricle	07.08.41
Single ventricle, Mitral atresia-modifier, Dominant right ventricle	07.08.42
Single ventricle, Tricuspid atresia	01.01.22, 06.01.01
Single ventricle, Tricuspid atresia (Right-sided atrioventricular valve atresia)	01.01.22, 06.01.01, 01.04.06
Single ventricle, Tricuspid atresia (Right-sided atrioventricular valve atresia), Type 1a {SDS} (No TGA with pulmonary atresia)	01.01.22, 01.03.00, 06.01.01, 01.04.06, 01.05.00, 09.05.11
Single ventricle, Tricuspid atresia (Right-sided atrioventricular valve atresia), Type 1b {SDS} (No TGA with pulmonary hypoplasia and with a small VSD)	01.01.22, 01.03.00, 06.01.01, 01.04.06, 01.05.00, 09.10.11, 07.14.01

Single ventricle, Tricuspid atresia (Right-sided atrioventricular valve atresia),Type 1c {SDS} (No TGA with no pulmonary hypoplasia and with a large VSD)	01.01.22, 01.03.00, 06.01.01, 01.04.06, 01.05.00, 09.10.11 + Q1.90.81, 07.14.04
Single ventricle, Tricuspid atresia (Right-sided atrioventricular valve atresia), Type 2a {SDD} (D-TGA with pulmonary atresia)	01.01.22, 01.03.00, 06.01.01, 01.04.06, 01.05.01, 09.05.11, 02.06.02
Single ventricle, Tricuspid atresia (Right-sided atrioventricular valve atresia), Type 2b {SDD} (D-TGA with pulmonary or subpulmonary stenosis)	01.01.22, 01.03.00, 06.01.01, 01.04.06, 01.05.01, 09.05.92, 02.06.02
Single ventricle, Tricuspid atresia (Right-sided atrioventricular valve atresia), Type 2c {SDD} (D-TGA with large pulmonary artery and unrestricted pulmonary blood flow)	01.01.22, 01.03.00, 06.01.01, 01.04.06, 01.05.01, 02.06.02, 09.05.94 + Q1.60.31
Single ventricle, Tricuspid atresia (Right-sided atrioventricular valve atresia), Type 3a {SDL} (L-TGA with pulmonary or subpulmonary stenosis)	01.01.22, 01.03.00, 06.01.01, 01.04.06, 01.05.01, 02.06.04, 09.05.92
Single ventricle, Tricuspid atresia (Right-sided atrioventricular valve atresia), Type 3b {SDL} (L-TGA with subaortic stenosis)	01.01.22, 01.03.00, 06.01.01, 01.04.06, 01.05.01, 02.06.04, 07.09.00
Single ventricle, Tricuspid atresia (Right-sided atrioventricular valve atresia), Type 3-With pulmonary atresia {SDL} (L-TGA with pulmonary atresia)	01.01.22, 01.03.00, 06.01.01, 01.04.06, 01.05.01, 02.06.04, 09.05.11
Single ventricle, Tricuspid atresia-Corrected transposition and left-sided atrioventricular valve atresia, {SLL}	**01.01.22, 01.03.00, 01.01.03, 06.01.02, 02.06.04, 02.03.02
Single ventricle, Tricuspid atresia-Corrected transposition and transleft-sided atrioventricular valve atresia, {SLL}, With pulmonary atresia	**01.01.22, 01.03.00, 01.01.03, 06.01.02, 02.06.04, 02.03.02, 09.05.11
Single ventricle, Tricuspid atresia-Corrected transposition and left-sided atrioventricular valve atresia, {SLL}, With pulmonary or subpulmonary stenosis	**01.01.22, 01.03.00, 01.01.03, 06.01.02, 02.06.04, 02.03.02, 09.05.92
Single ventricle, Tricuspid atresia-Corrected transposition and left-sided atrioventricular valve atresia, {SLL}, With subaortic stenosis	**01.01.22, 01.03.00, 01.01.03, 06.01.02, 02.06.04, 02.03.02, 07.09.00
Single ventricle, Tricuspid atresia-modifier, Dominant left ventricle	07.08.41
Single ventricle, Tricuspid atresia-modifier, Dominant right ventricle	07.08.42
Single ventricle, Unbalanced AV canal defect (Unbalanced atrioventricular septal defect)	01.01.22, 06.07.26
Single ventricle, Unbalanced AV canal defect (Unbalanced atrioventricular septal defect), Left dominant	06.07.06
Single ventricle, Unbalanced AV canal defect (Unbalanced atrioventricular septal defect), Right dominant	06.07.05
Single ventricle, Ventricular morphology uncertain	01.01.22, 07.08.44
Single ventricle, Ventricular morphology uncertain, Indeterminate ventricular morphology	01.01.22, 07.08.47
Single ventricle, Ventricular morphology uncertain, Mostly left ventricle	01.01.22, 07.08.45
Single ventricle, Ventricular morphology uncertain, Mostly right ventricle	01.01.22, 07.08.46

**The term congenitally corrected transposition is synonymous with discordant atrioventricular and ventriculo-arterial connections. The term can only be used in conjunction with tricuspid or mitral atresia if there is an imperforate atretic valve. In other cases of atresia, the atrioventricular connection is absent and therefore cannot be “discordant”, and the term congenitally corrected transposition does not apply

Table 2. The Association for European Paediatric Cardiology derived version of the International Paediatric and Congenital Cardiac Code for the Functionally Univentricular Heart.

Atrial arrangement (situs)	
Usual atrial arrangement (atrial situs solitus)	01.03.00
Mirror image atrial arrangement (atrial situs inversus)	01.03.01
Visceral heterotaxy (abnormal arrangement thoraco-abdominal organs)	03.01.02
Right isomerism ("asplenia")	03.01.04
Left isomerism ("polysplenia")	03.01.05
Double inlet ventricle	
Double inlet ventricle	01.01.14
Double inlet right ventricle	01.04.03
Double inlet left ventricle	01.04.04
Double inlet to solitary ventricle of indeterminate morphology	01.04.05
Two AV valves in double inlet ventricle	01.06.01
Common AV orifice in double inlet ventricle	01.06.02
Tricuspid atresia	
Tricuspid atresia	06.01.01
Absent right-sided AV connection (univentricular)	01.04.12
Absent right-sided AV connection with straddling valve (uniatrial biventricular)	01.04.15
Left-sided atrium to left ventricle	01.04.06
Left-sided atrium to right ventricle	01.04.07
Left-sided atrium to both ventricles	01.04.17
Left-sided atrium to ventricle of indeterminate morphology	01.04.08
Mitral Atresia	
Mitral atresia	06.02.01
Absent left-sided AV connection (univentricular)	01.04.13
Absent left-sided AV connection with straddling valve (uniatrial biventricular)	01.04.16
Right-sided atrium to right ventricle	01.04.09
Right-sided atrium to left ventricle	01.04.10
Right-sided atrium to both ventricles	01.04.18
Right-sided atrium to ventricle of indeterminate morphology	01.04.11
Ventriculo-arterial connection(s)	
Concordant VA connections	01.05.00
Discordant VA connections (TGA)	01.05.01
Double outlet right ventricle	01.01.04
Double outlet left ventricle	01.05.03
Common arterial trunk (truncus arteriosus)	09.01.01
Single outlet VA connection via aorta (pulmonary atresia)	01.05.32
Single outlet VA connection via pulmonary trunk (aortic atresia)	01.05.33
Solitary arterial trunk (absent intrapericardial pulmonary arteries)	09.07.26
Congenitally corrected transposition of great arteries (discordant AV & VA connections)	01.01.03
Ventricular topology	
Right hand pattern ventricular topology	02.03.01
Left hand pattern ventricular topology	02.03.02
Relationship of aortic orifice with respect to pulmonary orifice	
Aortic orifice posterior right with respect to pulmonary orifice (normal)	02.06.00
Aortic orifice right side-by-side with respect to pulmonary orifice	02.06.01
Aortic orifice anterior right with respect to pulmonary orifice	02.06.02
Aortic orifice anterior with respect to pulmonary orifice	02.06.03
Aortic orifice anterior left with respect to pulmonary orifice	02.06.04
Aortic orifice left side-by-side with respect to pulmonary orifice	02.06.05
Aortic orifice posterior left with respect to pulmonary orifice	02.06.06
Additional selected lesions and characteristics in functionally univentricular hearts	
Right ventricular hypoplasia	07.02.00
Left ventricular hypoplasia	07.07.00
Tricuspid valve atretic (imperforate)	06.01.02
Mitral valve atretic (imperforate)	06.02.02
Atrioventricular septal defect	06.06.00
AVSD with ventricular imbalance	06.07.26
AVSD with ventricular imbalance: dominant right ventricle, hypoplastic left ventricle	06.07.05
AVSD with ventricular imbalance: dominant left ventricle, hypoplastic right ventricle	06.07.06

(Continued)

Table 2. (Continued)

VSD	07.10.00
Restrictive VSD	07.14.01
Nonrestrictive VSD	07.14.04
Pulmonary stenosis	09.05.92
Pulmonary atresia	09.05.11
Pulmonary stenosis or atresia not present	09.05.94
Pulmonary arterial hypoplasia	09.10.11
Pulmonary arterial hypoplasia – not present	09.10.11 + Q1.90.81
Subpulmonary stenosis	07.05.30
Subpulmonary stenosis due to restrictive VSD in functionally univentricular heart	07.05.31
Subaortic stenosis	07.09.00
Subaortic stenosis due to restrictive VSD in functionally univentricular heart	07.09.18
– unrestricted pulmonary blood flow	Q1.60.31
– restricted pulmonary blood flow	Q1.60.32
– balanced systemic to pulmonary blood flow	Q1.60.33
Additional descriptors of functionally univentricular heart	
Functionally univentricular heart	01.01.22
Ventricular imbalance	07.08.40
Ventricular imbalance: dominant left ventricle + hypoplastic right ventricle	07.08.41
Ventricular imbalance: dominant right ventricle + hypoplastic left ventricle	07.08.42
Right ventricle not apparent	02.04.08
Left ventricle not apparent	02.04.09
Ventricular morphology uncertain	07.08.44
Ventricular morphology uncertain: probably dominant left ventricle	07.08.45
Ventricular morphology uncertain: probably dominant right ventricle	07.08.46
Ventricular morphology uncertain: probably ventricle of indeterminate morphology	07.08.47
Solitary ventricle of indeterminate morphology	02.03.05