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Author for correspondence:
Jeffrey P. Jacobs, MD, FACS, FACC, FCCP, Congenital Heart Center, UF Health Shands Hospital, Division of Cardiovascular Surgery, Departments of Surgery and Pediatrics, University of Florida, 1600 SW Archer Road, Gainesville, Florida, 32608, USA.
E-mails: jeffjacobs@msn.com and jeffreyjacobs@ufl.edu

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Nomenclature for Pediatric and Congenital Cardiac Care: Unification of Clinical and Administrative Nomenclature – The 2021 International Paediatric and Congenital Cardiac Code (IPCCC) and the Eleventh Revision of the International Classification of Diseases (ICD-11)

Jeffrey P. Jacobs[1], Rodney C. G. Franklin[2], Marie J. Béland[3], Diane E. Spicer[1,4], Steven D. Colan[5], Henry L. Walters III[6], Frédérique Bailliard[7,8], Lucile Houyel[9], James D. St. Louis[10], Leo Lopez[11], Vera D. Aiello[12], J. William Gaynor[13], Otto N. Krogmann[14], Hiromi Kurosawa[15], Bohdan J. Maruszewski[16], Giovanni Stellin[17], Paul Morris Weinberg[13], Marshall Lewis Jacobs[18], Jeffrey R. Boris[19], Meryl S. Cohen[13], Allen D. Everett[18], Jorge M. Giroud[20], Kristine J. Guleserian[21], Marina L. Hughes[22], Amy L. Juraszek[23], Stephen P. Seslar[24], Charles W. Shepard[25], Shubhika Srivastava[26], Andrew C. Cook[27], Adrian Crucean[28], Lazaro E. Hernandez[29], Rohit S. Loomba[30], Lindsay S. Rogers[13], Stephen P. Sanders[5], Jill J. Savla[13], Elif Seda Selamet Tierney[11], Justin T. Tretter[31], Lianyi Wang[32], Martin J. Elliott[33], Constantine Mavroudis[18,34] and Christo I. Tchervenkov[35]

[1]Congenital Heart Center, UF Health Shands Hospital, Division of Cardiovascular Surgery, Departments of Surgery and Pediatrics, University of Florida, Gainesville, FL, USA; [2]Paediatric Cardiology Department, Royal Brompton & Harefield NHS Trust, London, UK; [3]Division of Paediatric Cardiology, The Montreal Children's Hospital of the McGill University Health Centre, Montréal, Québec, Canada; [4]Johns Hopkins All Children's Hospital, Johns Hopkins University, Saint Petersburg, FL, USA; [5]Department of Cardiology, Boston Children's Hospital, Harvard University, Boston, MA, USA; [6]Cardiovascular Surgery, Children's Hospital of Michigan, Wayne State University School of Medicine, Detroit, MI, USA; [7]Bailliard Henry Pediatric Cardiology, Raleigh, North Carolina, United States of America; [8]Duke University, Durham, NC, USA; [9]Congenital and Pediatric Medico-Surgical Unit, Necker Hospital-M3C, Paris, France; [10]Department of Surgery and Pediatrics, Children Hospital of Georgia, Augusta University, Augusta, Georgia; [11]Lucile Packard Children's Hospital Stanford, Stanford University School of Medicine, Palo Alto, California, USA; [12]Heart Institute (InCor), University of São Paulo School of Medicine, São Paulo, Brazil; [13]Cardiac Center, Children's Hospital of Philadelphia, Philadelphia, PA, USA; [14]Pediatric Cardiology-Congenital Heart Disease, Heart Center Duisburg, Duisburg, Germany; [15]Cardiovascular Surgery, Tokyo Women's Medical University, Tokyo, Japan; [16]Department for Pediatric and Congenital Heart Surgery, Children's Memorial Health Institute, Warsaw, Poland; [17]Pediatric and Congenital Cardiac Surgical Unit, Department of Cardiothoracic and Vascular Sciences, University of Padova, Padova, Italy; [18]Johns Hopkins University, Baltimore, Maryland, USA; [19]Jeffrey R. Boris, MD LLC, Moylan, PA, USA; [20]All Children's Hospital, Saint Petersburg, FL, USA; [21]Congenital Heart Surgery, Medical City Children's Hospital, Dallas, TX, USA; [22]Cardiology Department, Norfolk and Norwich University Hospital NHS Trust, UK; [23]Terry Heart Institute, Wolfson Children's Hospital, Jacksonville, FL, USA; [24]Department of Pediatrics, Division of Pediatric Cardiology, Seattle Children's Hospital, University of Washington, Seattle, WA, USA; [25]Children's Heart Clinic of Minneapolis, Minneapolis, MN, USA; [26]Division of Cardiology, Department of Cardiovascular Medicine, Nemours Cardiac Center at the Alfred I. duPont Hospital for Children, Wilmington, DE, USA; [27]Institute of Cardiovascular Science, University College London, London, UK; [28]Congenital Heart Surgery, Birmingham Women's and Children's Foundation Trust Hospital, University of Birmingham, Birmingham, UK; [29]Joe DiMaggio Children's Hospital Heart Institute, Hollywood, FL, USA; [30]Advocate Children's Heart Institute, Advocate Children's Hospital, Oak Lawn, IL, USA; [31]Department of Pediatrics, Heart Institute, Cincinnati Children's Hospital Medical Center, University of Cincinnati College of Medicine, Cincinnati, OH, USA; [32]Heart Centre, First Hospital of Tsinghua University, Beijing, China; [33]University College London, London, UK; [34]Peyton Manning Children's Hospital, Indianapolis, IN, USA and [35]Division of Cardiovascular Surgery, The Montreal Children's Hospital of the McGill University Health Centre, Montréal, Québec, Canada

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Abstract

Substantial progress has been made in the standardization of nomenclature for paediatric and congenital cardiac care. In 1936, Maude Abbott published her Atlas of Congenital Cardiac Disease, which was the first formal attempt to classify congenital heart disease. ***The International Paediatric and Congenital Cardiac Code*** (***IPCCC***) is now utilized worldwide and has most recently become the paediatric and congenital cardiac component of the ***Eleventh Revision of the International Classification of Diseases*** (***ICD-11***). The most recent publication of the ***IPCCC*** was in 2017. This manuscript provides an updated 2021 version of the ***IPCCC***.

The International Society for Nomenclature of Paediatric and Congenital Heart Disease (***ISNPCHD***), in collaboration with the ***World Health Organization (WHO***), developed the paediatric and congenital cardiac nomenclature that is now within the eleventh version of the International Classification of Diseases (ICD-11). This unification of ***IPCCC*** and ICD-11 is the ***IPCCC ICD-11 Nomenclature*** and is the first time that the clinical nomenclature for paediatric and congenital cardiac care and the administrative nomenclature for paediatric and congenital cardiac care are harmonized. The resultant congenital cardiac component of ICD-11 was increased from 29 congenital cardiac codes in ICD-9 and 73 congenital cardiac codes in ICD-10 to 318 codes submitted by ***ISNPCHD*** through 2018 for incorporation into ICD-11. After these 318 terms were incorporated into ICD-11 in 2018, the ***WHO*** ICD-11 team added an additional 49 terms, some of which are acceptable legacy terms from ICD-10, while others provide greater granularity than the ***ISNPCHD*** thought was originally acceptable. Thus, the total number of paediatric and congenital cardiac terms in ICD-11 is 367. In this manuscript, we describe and review the terminology, hierarchy, and definitions of the ***IPCCC ICD-11 Nomenclature***. This article, therefore, presents a global system of nomenclature for paediatric and congenital cardiac care that unifies clinical and administrative nomenclature.

The members of ***ISNPCHD*** realize that the nomenclature published in this manuscript will continue to evolve. The version of the ***IPCCC*** that was published in 2017 has evolved and changed, and it is now replaced by this 2021 version. In the future, ***ISNPCHD*** will again publish updated versions of ***IPCCC***, as ***IPCCC*** continues to evolve.

Introduction

Substantial progress has been made in the standardization of nomenclature for paediatric and congenital cardiac care.[1–108] In 1936, Maude Abbott, of McGill University in Montréal, Québec, Canada, published her Atlas of Congenital Cardiac Disease, which was the first formal attempt to classify congenital heart disease.[1] ***The International Paediatric and Congenital Cardiac Code*** (***IPCCC***) is now utilized worldwide and is the paediatric and congenital cardiac component of the ***Eleventh Revision of the International Classification of Diseases*** (***ICD-11***).[105,106] The most recent publication of the ***IPCCC*** was in 2017.[105] This manuscript provides an updated 2021 version of the ***IPCCC***, which is now the paediatric and congenital cardiac component of ***ICD-11***.

Congenital cardiac malformations are the most common types of birth defects. Before the introduction of current diagnostic modalities, such as echocardiography, the estimated incidence of CHD ranged from five to eight per 1000 live births. With improved diagnostic modalities, many more patients with milder forms of CHD can now be identified, so that contemporary estimates of the prevalence of congenital cardiac disease now range from eight to twelve per 1000 live births.[109–112]. About one-quarter of neonates and infants with a congenital cardiac defect undergo surgery or catheter-directed intervention in their first year of life.[109] Survival after surgery for congenital heart defects has increased over the past decade, especially for the most complex operations.[113] The aetiology of this improvement is obviously multifactorial, but the ability to compare and benchmark risk-stratified and risk-adjusted outcomes at individual programs to national and international aggregate benchmarks has certainly facilitated these improved cardiac surgical outcomes over time. This benchmarking and improvement in quality requires standardization of the nomenclature and classification of paediatric and congenital cardiac disease, as described in this manuscript.

This manuscript presents the latest edition of ***The International Paediatric and Congenital Cardiac Code*** (***IPCCC***), which has been integrated into the paediatric and congenital cardiac component of the ***Eleventh Revision of the International Classification of Diseases*** (***ICD-11***). This article will discuss the following topics:

- **The International Paediatric and Congenital Cardiac Code (IPCCC)**
- **The Eleventh Revision of the International Classification of Diseases (ICD-11)**
- **Clinical Nomenclature versus Administrative Nomenclature**

This article will then present the following three **Tables of IPCCC ICD-11 Nomenclature for Congenital Cardiac Diagnostic Terms in the *ICD-11 Foundation***

- **Table 1. IPCCC ICD-11 Diagnostic Hierarchy**
- **Table 2. IPCCC ICD-11 Definitions**
- **Table 3. IPCCC ICD-11 Codes**

The version of the ***IPCCC*** that was published by ***ISNPCHD*** in 2017[105] has evolved and is now updated with the 2021 version published in this manuscript. In the future, ***ISNPCHD*** will again publish additional updated versions of ***IPCCC***, as ***IPCCC*** continues to evolve.

The International Paediatric and Congenital Cardiac Code (IPCCC)

As already emphasised, the development of classification schemes specific to the congenitally malformed heart began with Maude Abbott's pioneering work in the early 1900s.[1,105] Her landmark publication in 1936, entitled "Atlas of Congenital Cardiac Disease", was the first formal attempt to classify the lesions seen when the heart is congenitally malformed.[1,105] It was not until the 1990s that efforts were made to create a truly international system of nomenclature and classification to support paediatric and congenital cardiac care. Prior to these efforts of the 1990s, multiple systems of nomenclature and classification were used at hospitals across the world. These various systems of nomenclature were the basis of internal, national, and even international registries and databases of paediatric and congenital cardiac care.[105]

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Aided by advances in information technology that facilitate the exchange of information, two independent international collaborations began in the 1990s, resulting in the publication of two separate international paediatric and congenital cardiac systems of nomenclature and classification:

- *The European Paediatric Cardiac Code* (*EPCC*) of *The Association for European Paediatric and Congenital Cardiology* (*AEPC*)[2,3]
- The nomenclature system of the *International Congenital Heart Surgery Nomenclature and Database Project* of *The Society of Thoracic Surgeons* (*STS*) in North America, *The European Association for Cardio-Thoracic Surgery* (*EACTS*), and The European Congenital Heart Defects Database of *The European Congenital Heart Surgeons Foundation* (*ECHSF*) – (renamed *The European Congenital Heart Surgeons Association* [*ECHSA*] in 2003).[4–37]

During the 1990s, both ECHSF and STS created databases to assess the outcomes of congenital cardiac surgery. Beginning in 1998, EACTS, ECHSA, and STS collaborated to create the International Congenital Heart Surgery Nomenclature and Database Project. As a result of this project, by 2000, a common nomenclature, along with a common core minimal dataset, were adopted by EACTS, ECHSA, and STS, and published in ***The Annals of Thoracic Surgery*** as a 372-page free standing Supplement.[4–37] In parallel, in 1996, the AEPC created a Coding Committee to produce a set of diagnostic and procedural codes that would be acceptable and adopted within both the European paediatric cardiology and European paediatric cardiac surgical communities. As a result of this project, in 2000, the EPCC was published in ***Cardiology in the Young*** as a 146-page free standing Supplement.[2,3]

Both the EPCC and the International Congenital Heart Surgery Nomenclature and Database Project included a comprehensive Long List, with thousands of terms, and a Short List designed to be used as part of a minimum data set for multi-institutional registries and databases. Both Long Lists mapped fully to their respective Short Lists. The nearly simultaneous publication of these two complementary systems of nomenclature led to the problematic situation of having two systems of nomenclature that were to be widely adopted, with the potential risk of duplicate or inaccurate coding within institutions, as well as the potential problem of invalidating multicentric projects owing to confusion between the two systems.[105]

Hence, on Friday, October 6, 2000, in Frankfurt, Germany, during the meeting of ECHSF prior to the 14th Annual Meeting of EACTS, representatives of the involved Societies met and established ***The International Nomenclature Committee for Paediatric and Congenital Heart Disease***, which was to include representatives of the four societies (AEPC, STS, ECHSF, and EACTS), as well as representatives from the remaining continents of the world – Africa, Asia, Australia (Oceania), and South America.[41–45,47,105] Over four years later, in January, 2005, ***The International Nomenclature Committee for Pediatric and Congenital Heart Disease*** was constituted and legally incorporated as ***The International Society for Nomenclature of Paediatric and Congenital Heart Disease*** (***ISNPCHD***).

At the meeting in Frankfurt in 2000, an agreement was reached to collaborate and produce a reconciliatory bidirectional map between the two systems of nomenclature. The feasibility of this project was established by the creation of a rule-based bidirectional crossmap between the two Short Lists, using the six-digit coding system already established within the EPCC as the common link between the two nomenclature lexicons. This bidirectional crossmap between the two Short Lists was created and published by ***The International Working Group for Mapping and Coding of Nomenclatures for Pediatric and Congenital Heart Disease***, also known for short as the ***Nomenclature Working Group*** (***NWG***), which was the original committee of ***The International Nomenclature Committee for Paediatric and Congenital Heart Disease*** and the subsequent ***ISNPCHD***[44,45,47]

Over the next 8 years, the NWG met 10 times, over a combined period of 47 days, to achieve the main goal of crossmapping the two comprehensive Long Lists to create the IPCCC, which has two dominant versions[105]:

- The version of the IPCCC derived from the European Paediatric Cardiac Code of AEPC
- The version of the IPCCC derived from the International Congenital Heart Surgery Nomenclature and Database Project of EACTS, ECHSA, and STS.

These two versions of the IPCCC are crossmapped to each other by means of the six-digit coding system[63] and have the following abbreviated short names:

- EACTS-STS derived version of the IPCCC
- AEPC derived version of the IPCCC

The NWG therefore crossmapped the nomenclature of the International Congenital Heart Surgery Nomenclature and Database Project of EACTS, ECHSA, and STS with the EPCC of AEPC, and thus created the IPCCC,[63] which is available for free download from the internet at [https://www.IPCCC.net]. Additional systems of nomenclature, for paediatric cardiology and cardiac surgery, which were mapped to the common six-digit code spine, include the Boston-based Fyler codes, and the Canadian nomenclature system. There is also mapping to the ninth and tenth revisions of the International Classification of Diseases (ICD-9, ICD-10), usually in a many to one fashion, given the limitations of these earlier versions of ICD.

Most international databases of patients with paediatric and congenital cardiac disease now use the IPCCC as their foundation. This common nomenclature, the IPCCC, and the common minimum database data set created by the International Congenital Heart Surgery Nomenclature and Database Project, are now utilized by multiple databases and registries of paediatric and congenital cardiac care across the world. The following databases all use the EACTS-STS derived version of the IPCCC:

- The Society of Thoracic Surgeons Congenital Heart Surgery Database (STS CHSD),[39,40,46]
- The European Congenital Heart Surgeons Association Congenital Heart Surgery Database (ECHSA CHSD),[39,40,46]
- The Japan Congenital Cardiovascular Surgery Database (JCCVSD),[39,40,46] and
- The World Database for Pediatric and Congenital Heart Surgery (WDPCHS).[114]

Several national and institutional databases in Europe use the AEPC derived version of the IPCCC for collection of data, including:

- Germany,
- the Netherlands, and

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- the United Kingdom and Republic of Ireland National Congenital Heart Disease Audit.

For all terms within the two versions of the IPCCC, a unique six-digit code corresponds to a single entity, whether it be a morphological phenotype, procedure, symptom, or genetic syndrome.[105] The mapped terms in each of the two versions are synonymous.[105] By 2013, there were 12,168 terms in the IPCCC Long List version derived from the European Paediatric Cardiac Code, and 17,176 terms in the IPCCC Long List version derived from the International Congenital Heart Surgery and Nomenclature Database Project. These Long Lists include hundreds of qualifiers, some specific, such as anatomical sites, and others generic, such as gradings of severity.

It is primarily the Short Lists, rather than the Long Lists, of the two crossmapped versions of the IPCCC that have been used for analyses of multi-institutional and international outcomes following operations and procedures for patients with congenitally malformed hearts. Over a million patients are now coded with the IPCCC in registries worldwide.[105] Both versions of the IPCCC Short Lists have been used to develop empirical systems for the adjustment of risk following surgical procedures, based on the operation type and comorbidities, for the purposes of quality assurance and quality improvement.[115–119] Both risk adjustment systems depend upon the IPCCC for all variables, to ensure a common nomenclature between institutions submitting data, and both perform better than the systems based on the subjective assessment of risk.[105]

The history of ***ISNPCHD*** and the development of ***IPCCC*** have been previously published.[44,45,47,105,107,108] The International Working Group for Mapping and Coding of Nomenclature for Paediatric and Congenital Heart Disease was also known as the Nomenclature Working Group or NWG and was the first committee of ***The International Society for Nomenclature of Paediatric and Congenital Heart Disease*** (***ISNPCHD***). The initial 12 members of Nomenclature Working Group represented multiple subspecialties and continents:

- Vera Aiello, University of São Paulo Medical School, São Paulo, Brazil
- Marie J. Béland, The Montreal Children's Hospital, Montréal, Québec, Canada
- Steven Colan, Boston Children's Hospital, Boston, Massachusetts, United States of America
- Rodney C. G. Franklin, Royal Brompton & Harefield Hospital NHS Foundation Trust, London, United Kingdom
- J. William Gaynor, The Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, United States of America
- Jeffrey P. Jacobs, University of Florida, Gainesville, Florida, United States of America
- Otto N. Krogmann, Heart Center Duisburg, Duisburg, Germany
- Hiromi Kurosawa, Tokyo Women's Medical University, Tokyo, Japan
- Bohdan J. Maruszewski, Children's Memorial Health Institute, Warsaw, Poland
- Giovanni Stellin, Universita di Padova, Italy
- Christo I. Tchervenkov, The Montreal Children's Hospital, Montréal, Québec, Canada
- Paul Weinberg, The Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, United States of America

The Presidents of ***The International Society for Nomenclature of Paediatric and Congenital Heart Disease*** (***ISNPCHD***) are listed below, along with the terms of their Presidency:

- Martin J. Elliott (2000–2009)
- Christo I. Tchervenkov (2009–2013)
- Rodney C. G. Franklin (2013–2017)
- Jeffrey P. Jacobs (2017–2021)
- Steven D. Colan (2021–2025)

Although both the Short Lists and the comprehensive Long Lists of each version of IPCCC have been crossmapped, the two Short Lists emanating from their respective Long-List versions are not the same in terms of structure or content.[105] ***ISNPCHD*** recognized this disparity, and believed that the creation of a congenital cardiac subset within ICD-11 would accomplish several goals:

- help resolve the differences between the Short List of the EACTS-STS derived version of the IPCCC and the Short List of the AEPC derived version of the IPCCC
- present a single common comprehensive and hierarchical Short List of diagnostic terms that could serve all communities involved with paediatric and congenital cardiac care
- harmonize the administrative nomenclature for paediatric and congenital cardiac care with the clinical nomenclature for paediatric and congenital cardiac care.

Hence, ***ISNPCHD*** created, organized, and defined the terms of IPCCC in order to standardize nomenclature for paediatric and congenital cardiac care and promote accurate coding, sharing of information, and analysis of data.[41–45,47,105] ***ISNPCHD*** believed from the start that the concept of "illustration" of the terms would be very important to advance these goals.[87–90,93,94,102] Concurrent with its involvement in developing ICD-11, as described in detail below, ***ISNPCHD*** began creating the ***IPCCC ICD-11 Congenital Heart Atlas*** to illustrate the terms listed in the "Structural developmental anomaly of heart or great vessels" section of ICD-11. In addition to the terms, definitions, and data about coding that is published in ICD-11, the ***IPCCC ICD-11 Congenital Heart Atlas*** is currently being built to contain drawings, photographs of anatomical specimens, images and videos from various imaging modalities, and intraoperative photographs and videos, all designed to help health care professionals better select the correct designation for the cardiac phenotypes listed in ICD-11. The ***IPCCC ICD-11 Congenital Heart Atlas*** will, of course, also fulfill multiple educational purposes. The ***IPCCC ICD-11 Congenital Heart Atlas*** will be freely accessible on the ***ISNPCHD*** website: [https://www.IPCCC.net]. The ***IPCCC ICD-11 Congenital Heart Atlas*** will also be freely accessible via hyperlinks from:

- Heart University [https://www.heartuniversity.org/], and
- The World University for Pediatric and Congenital Heart Surgery [https://www.wupchs.education]

The Eleventh Revision of the International Classification of Diseases (ICD-11)

The history of ***The International Classification of Diseases*** (***ICD***) dates back to the late 1800s (Figure 1):

- In 1891, the International Statistical Institute commissioned a committee chaired by Jacques Bertillon (1851–1922), Chief of

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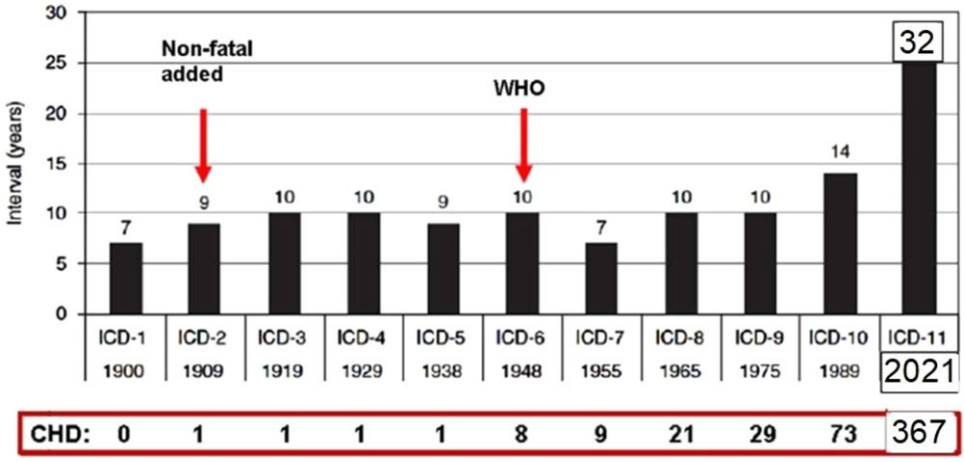

Figure 1. *The International Classification of Diseases* (*ICD*). This bar chart documents the time interval between each Revision of the International Classification of Diseases (ICD), 1900–2021. The horizontal lower bar indicates the number of terms related to congenital heart disease (CHD) listed in each ICD version.

Statistical Services of the City of Paris, to create what became the Bertillon [International] Classification of Causes of Death, with associated sequential numeric codes.[105] Over the following decades, this classification scheme was adopted by many countries in the Americas and Europe, with conferences for revision occurring roughly every 10 years to update the system, which became known as ***The International Classification of Diseases*** (***ICD***).

- In 1893, Bertillon presented the (International) Classification of Causes of Death at the meeting of the International Statistical Institute in Chicago, where it was adopted by several cities and countries.
- In 1898, the American Public Health Association recommended its adoption in North America, and that the classification be revised every 10 years.
- In 1900, the First International Conference to revise the Bertillon Classification of Causes of Death was held in Paris.
- In 1909, non-fatal diseases, in other words, morbidity, were added.
- From 1948 until now, the ***World Health Organization (WHO)*** has promoted and managed ICD, starting in 1948 with the sixth revision of the International Classification of Diseases, Injuries and Causes of Death.

According to ***WHO***, "ICD is the foundation for the identification of health trends and statistics globally, and the international standard for reporting diseases and health conditions. It is the diagnostic classification standard for all clinical and research purposes. ICD defines the universe of diseases, disorders, injuries and other related health conditions, listed in a comprehensive, hierarchical fashion" [https://www.who.int/standards/classifications/classification-of-diseases]. The ICD-11 development mission was "To produce an international disease classification that is ready for electronic health records that will serve as a standard for scientific comparability and communication".[105] ICD-11 was officially launched on-line by the ***WHO*** in June 2018 and endorsed by the World Health Assembly on 25 May 2019. The ***WHO*** states that ICD-11 is to be "The global standard for health data, clinical documentation, and statistical aggregation", that it is "scientifically up-to-date and designed for use in the digital world with state-of-the art technology to reduce the costs of training and implementation", and that its "multilingual design facilitates global use" [https://www.who.int/classifications/classification-of-diseases]. The purpose of ICD-11 "is to allow the systematic recording, analysis, interpretation, and comparison of mortality and morbidity data collected in different countries or areas and at different times".[105] The ICD-11 project began in earnest in 2007. Importantly, ICD-11 incorporates textual definitions. With the creation of ICD-11, for the first time, the revision process moved away from reliance on large meetings of national delegations of health statisticians, wherein those who voiced their opinion strongest would dominate the content of the paper-based output – "decibel" diplomacy. In contrast, the ICD-11 revision process is dependent upon international expert clinicians, with digital curation, the incorporation of wide peer review, and extensive field testing. "ICD-11 has been adopted by the Seventy-second World Health Assembly in May 2019 and comes into effect on 1 January 2022" [https://www.who.int/standards/classifications/classification-of-diseases].

The task of creating ICD-11 was divided into content specific Topic Advisory Groups, with related Working Groups led by Managing Editors and chaired by specialist clinicians with an intentionally wide geographic spread. From 2009 through to 2016, the Managing Editor coordinated a series of meetings, some face-to-face, but mostly teleconferences, beginning with the hierarchical structure and terms within ICD-10, and initially producing an evolving alpha draft. In 2012, a beta draft was published online [https://icd.who.int/dev11/f/en], coinciding with the authoring process moving to a web-based platform for its entire content.[105] The tool allows online global peer review and submission of comments by both the authors and worldwide interested parties in the field-testing stage.

From the start, clinicians involved in the Topic Advisory Groups have been encouraged to enlist the advice of specialist Societies to aid the process, thus ensuring that the content was both up-to-date and had Societal endorsement. This process has resulted in a huge increase in the number of individual terms within ICD-11, with secondary expansion of the hierarchical structure when compared with ICD-10.

In collaboration with ***WHO***, ***ISNPCHD*** developed the paediatric and congenital cardiac nomenclature that is now within the eleventh version of the International Classification of Diseases (ICD-11).[105] This unification of IPCCC and ICD-11 is the ***IPCCC ICD-11 Nomenclature*** and is the first time that the clinical nomenclature for paediatric and congenital cardiac care and

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the administrative nomenclature for paediatric and congenital cardiac care have been harmonized.[105] The resultant congenital cardiac component of ICD-11 was increased from 29 CHD diagnostic terms codes in ICD-9 and 73 CHD diagnostic terms in ICD-10 to 318 codes submitted by ***ISNPCHD*** through 2018 for incorporation into ICD-11.[105] After these 318 terms were incorporated into ICD-11 in 2018, the ***WHO*** ICD-11 team added an additional 49 terms, some of which are acceptable legacy terms from ICD-10, while others provide greater granularity than the ***ISNPCHD*** thought was originally acceptable, such as individual codes for the various types of isolated branches of the aortic arch or branches of the aortic arch having an aberrant origin. Thus, the total number of paediatric and congenital cardiac terms in ICD-11 is now 367. (Tables 1–3). Populating ICD-11 by the content-specific Topic Advisory Groups was not always without controversy, with at times, for example, heated and prolonged discussions between the Rare Diseases Topic Advisory Group and several Internal Medicine Topic Advisory Workings Groups, including the Cardiovascular Working Group, over the hierarchy and content to be included or excluded. Tables 1 and 2 present the diagnostic hierarchy (Table 1) and definitions (Table 2) of the 318 codes submitted by ***ISNPCHD*** to compose the **IPCCC ICD-11 Nomenclature**, as well as the additional 49 scientifically correct or legacy terms added by the ***WHO*** ICD-11 team. As these additional 49 entities have now been added to IPCCC, ***ISNPCHD*** has provided the needed definitions for these terms (as presented in Table 2). Other legacy and scientifically incorrect terms inserted into the ***ICD-11 Foundation*** by the ***WHO*** ICD-11 team were judged by ***ISNPCHD*** to be obsolete or meaningless. These obsolete or meaningless terms, such as "Transposition of the aorta" and "Accessory heart", have been highlighted to ***WHO*** and have been made 'obsolete' within the system, meaning that these terms are retained for legacy purposes but will not be visible nor easily searchable. Tables 1–3, therefore, present the 367 terms that are part of IPCCC and also the paediatric and congenital cardiac component of ICD-11. Consequently, IPCCC and ICD-11 are a system of nomenclature that will, for the first time ever, harmonize the administrative nomenclature for paediatric and congenital cardiac care with the clinical nomenclature for paediatric and congenital cardiac care. This important goal will be achieved with the implementation of ICD-11.

Another of the aims of WHO for ICD-11 is to have the entirety of ICD-11 translated into different languages. The achievement of this objective will enhance the global uptake and utility of ICD-11 for international comparisons of outcomes and initiatives of quality improvement. Currently WHO list 22 languages which are at least partially complete. Knowing this fact, members of ***ISNPCHD*** have already translated the IPCCC ICD-11 Nomenclature into French[120] and Portuguese.[121] ***ISNPCHD*** has submitted the French version into ICD-11 via their translation tool platform. Unfortunately, it has become apparent that much of the translation work has been delegated by WHO to national governmental designated translation teams, without input from clinicians. This suboptimal strategy has led to some clinically unusable translations in the field of congenital cardiac care in ICD-11. For example:

- English IPCCC term currently in ICD-11: Double outlet right ventricle with non-committed ventricular septal defect
- ISNPCHD French translation: Ventricule droit à double issue avec communication interventriculaire sans relation avec les deux gros vaisseaux
- WHO translation (done without ISNPCHD input): Ventricule droit à double sortie avec anomalie septale ventriculaire à distance
- English equivalent to WHO translation: *Double exit right ventricle with ventricular septal anomaly at a distance*

An anglophone clinician would probably understand what is meant by Double exit right ventricle", but clearly "ventricular septal anomaly at a distance" does not convey the same information as the phrase "non-committed ventricular septal defect". This suboptimal translation and other similar errors need to be corrected. Fortunately, WHO have recently agreed to facilitate members of the ISNPCHD French translation team to work with the French government translation team to resolve these important issues.

The Foundation Component of ICD-11 (ICD-11 Foundation)

The full ICD-11 content is known as the ***ICD-11 Foundation***, which represents the entire ICD-11 universe, divided into 26 sections, and can be accessed digitally [https://icd.who.int/dev11/f/en#/http%3a%2f%2fid.who.int%2ficd%2fentity%2f455013390]. The 318 diagnostic terms for CHD that were submitted by ***ISNPCHD*** in 2018 reside in the **Foundation Component of ICD-11**, within the **Developmental Anomalies section**, with the parent term "**Structural developmental anomaly of heart or great vessels**", along with the additional 49 terms added to IPCCC by the ***WHO*** ICD-11 team since 2018.

The ICD-11 Mortality and Morbidity Statistics version (ICD-11 MMS)

Another feature of ICD-11 is that it is designed to be explicitly stratified to cater to different users, such as primary care, traditional medicine, and public health, producing so-called linearizations or "Tabular Lists". The initial and most important overall linearization of ICD-11 was that published in July 2018 as the ***Mortality and Morbidity Statistics version***, known as ***ICD-11-MMS***, with a 'blue' website: [https://icd.who.int/browse11/l-m/en], which is separate from the 'orange' ***ICD-11 Foundation*** website: [https://icd.who.int/dev11/f/en#/http%3a%2f%2fid.who.int%2ficd%2fentity%2f455013390].

ICD-11-MMS is the nearest equivalent to previous ICD versions. ***ICD-11-MMS*** includes a printed copy of top-level terms, and is designed to collect global data at a level of detail sufficient to capture important trends in the causes of death and prevalence of major disease entities. It is also the likely diagnostic coding system that will be used by nations for billing purposes. To achieve this objective, ***WHO*** in effect top-sliced the ***ICD-11 Foundation*** level content to include relevant higher-level terms, although not always with the input of clinicians. In addition, and consistent with previous ICD versions, the ***WHO*** has added two additional generic terms in each subsection of the ***ICD-11-MMS***:

1. "Other specified … disease" (Y-codes). For example: *LA89.Y Other specified functionally univentricular heart*
2. "Disease …, unspecified" (Z-codes), which are equivalent to Not Otherwise Specified (NOS) in previous ICD versions. For example: *LA89.Z Functionally univentricular heart, unspecified*. Of note is that LA89 itself is the MMS code for *Functionally univentricular heart*.

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Another example of the Y and Z codes is provided below:

1. Other specified . . . disease" (Y-codes). For example: *LA87.0Y Other specified anomaly of tricuspid valve*
2. "Disease . . . , unspecified" (Z-codes). For example: *LA87.0Z Congenital anomaly of tricuspid valve, unspecified*. Of note is that LA87.0 itself is the MMS code for Congenital anomaly of tricuspid valve.

These Y and Z codes do not appear in the ***ICD-11 Foundation***. Y and Z codes are unique to the ***ICD-11-MMS*** version, as will be described in the following discussion. For example, the term "Straddling tricuspid valve" can be found in ***ICD-11 Foundation***, but is not listed in ***ICD-11 MMS***. If coding with ***ICD-11 MMS***, the code LA87.0Y should be used to indicate that a more specific diagnosis is known.

For CHD, of the 367 paediatric and congenital cardiac terms currently in the ***ICD-11 Foundation***, a subset of 104 terms have been retained and will appear in the ***ICD-11-MMS*** linearization. As the ***ICD-11-MMS*** is likely to be the first component of ICD-11 to be adopted by countries worldwide, ***ISNPCHD*** has created a many-to-one unidirectional map of the CHD ***ICD-11 Foundation*** level content to the CHD ***ICD-11-MMS*** content within Developmental Anomalies (Chapter 20). This many-to-one unidirectional map of the CHD ***ICD-11 Foundation*** level content to the anticipated 2022 version of the CHD ***ICD-11-MMS*** is provided in Table 3 of this manuscript.

Clinical nomenclature versus administrative nomenclature

Several studies have examined the relative utility of clinical and administrative nomenclature for the evaluation of quality of care for patients undergoing treatment for paediatric and congenital cardiac disease. Evidence from four investigations suggests that the validity of coding of lesions seen in the congenitally malformed heart via the 9th Revision of the International Classification of Diseases (ICD-9) is poor.[65,91,122,123]

- First, in a series of 373 infants with congenital cardiac defects at Children's Hospital of Wisconsin, investigators reported that only 52% of the cardiac diagnoses in the medical records had a corresponding code from the ICD-9 in the hospital discharge database.[122]
- Second, the Hennepin County Medical Center discharge database in Minnesota identified all infants born during 2001 with a code for congenital cardiac disease using ICD-9. A review of these 66 medical records by physicians was able to confirm only 41% of the codes contained in the administrative database from ICD-9.[123]
- Third, the Metropolitan Atlanta Congenital Defect Program of the Birth Defect Branch of the Centers for Disease Control and Prevention of the United States government carried out surveillance of infants and fetuses with cardiac defects delivered to mothers residing in Atlanta during the years 1988 through 2003.[65] These records were reviewed and classified using both administrative coding and the clinical nomenclature used in the Society of Thoracic Surgeons Congenital Heart Surgery Database. This study concluded that analyses based on the codes available in ICD-9 are likely to "have substantial misclassification" of congenital cardiac disease.
- Fourth, a study was performed using linked patient data (2004-2010) from the Society of Thoracic Surgeons Congenital Heart Surgery (STS-CHS) Database (clinical registry) and the Pediatric Health Information Systems (PHIS) database (administrative database) from hospitals participating in both in order to evaluate differential coding/classification of operations between datasets and subsequent impact on outcomes assessment.[91] The cohort included 59,820 patients from 33 centres. There was a greater than 10% difference in the number of cases identified between data sources for half of the benchmark operations. The negative predictive value (NPV) of the administrative (versus clinical) data was high (98.8%-99.9%); the positive predictive value (PPV) was lower (56.7%-88.0%). These differences translated into significant differences in outcomes assessment, ranging from an underestimation of mortality associated with truncus arteriosus repair by 25.7% in the administrative versus clinical data (7.01% versus 9.43%; $p = 0.001$) to an overestimation of mortality associated with ventricular septal defect (VSD) repair by 31.0% (0.78% versus 0.60%; $p = 0.1$). This study demonstrates differences in case ascertainment between administrative and clinical registry data for children undergoing cardiac operations, which translated into important differences in outcomes assessment.

As discussed below, these challenges and problems persist with the 10th Revision of the International Classification of Diseases (ICD-10). Several potential reasons can explain the poor diagnostic accuracy of administrative databases and codes from ICD-9 and even ICD-10:

- accidental miscoding;
- coding performed by medical records clerks who have never seen the actual patient, in other words, coding performed by personnel not involved in the care of the patient;
- contradictory or poorly described information in the medical record;
- lack of diagnostic specificity for congenital cardiac disease in the codes of ICD-9 or ICD-10
- inadequately trained medical coders.

Although one might anticipate some improvement in diagnostic specificity with the adoption of ICD-10, it is still substantially deficient compared to that currently achieved with the clinical nomenclature used in clinical registries. In this regard, ICD-9 has only 29 congenital cardiac codes while ICD-10 has only 73 congenital cardiac codes. It will not be until there is implementation of the paediatric and congenital cardiac components of ICD-11 that harmonization of clinical and administrative nomenclature will be achieved. The implementation of ICD-11, therefore, will resolve many of these challenging issues.

Summary

The art and science of outcomes analysis and quality improvement for paediatric and congenital cardiac care continue to evolve. The IPCCC nomenclature is utilized in multi-institutional registries and databases all over the world.[124,125] In this manuscript, we have presented the 2021 version of IPCCC, a global system of nomenclature for paediatric and congenital cardiac care that unifies clinical and administrative nomenclature.

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Tables of IPCCC ICD-11 nomenclature for congenital cardiac diagnostic terms in *ICD-11 Foundation*

- Table 1 presents the diagnostic hierarchy of the paediatric and congenital cardiac terms in the ***ICD-11 Foundation***. Terms that appear in the ***ICD-11 MMS*** are presented in rows highlighted in yellow.
- Table 2 contains the definitions, commentary, synonyms, and abbreviations for these terms of the paediatric and congenital cardiac terms in the ***ICD-11 Foundation***. Terms that appear in the ***ICD-11 MMS*** are presented in rows highlighted in yellow.
- Table 3 contains the various IPCCC ICD-11 Codes, including the IPCCC codes as well as the ICD-11 Foundation entity numbers and the ICD-11 MMS codes.

In the Tables:

$^{+}$ = New terms added by the ***WHO*** ICD-11 team since the original 318 terms contained in the publication[105] from 2017
** = Terms that code normal human anatomy, but are important to specify when part of a complex congenital cardiac malformation
*** = Terms that are not located in the paediatric and congenital cardiac section of ICD-11
Rows with numbers in the second column labelled "ICD-11 Row Number or Letter" contain terms in the original 318 terms contained in the publication[105] from 2017.
Rows with letters in the second column labelled "ICD-11 Row Number or Letter" contain new terms added by the ***WHO*** ICD-11 team since the original 318 terms contained in the publication[105] from 2017.

MMS coding notes for Table 3:

1) The column titled "**ICD-11 MMS code or ICD-11 MMS crossmap**" contains the alphanumeric codes for terms listed in ICD-11 Mortality and Morbidity Statistics (highlighted in yellow). For the terms that are not highlighted (not in ICD-11 MMS), the column contains the alphanumeric codes of higher order MMS terms to which they have been cross-mapped. For example, "Complete agenesis of pericardium" (not listed in MMS), has been crossmapped to MMS term "Congenital pericardial anomaly" (LA8D).
2) Several terms in MMS have two additional versions (not published here) with distinct alphanumeric codes (either ending in "Y" [Other specified] or "Z" [unspecified]). For example, in addition to the term "Congenital anomaly of coronary artery" (LA8C), the following terms also exist: "Other specified congenital anomaly of coronary artery" (LA8C.Y) and "Congenital anomaly of coronary artery, unspecified" (LA8C.Z). When available, the non-highlighted terms in Table 3 have been crossmapped to the "Y" version of higher order MMS terms, since it conveys the added information that a more specific diagnosis is known, but that the more specific term does not exist in MMS. For example, "Accessory coronary artery" has been crossmapped to LA8C.Y instead of LA8C.
3) The first term of Table 3, "Structural developmental anomaly of heart or great vessels", is listed in MMS but does not have an MMS alphanumeric code. If wanting to code for this term in MMS, one must use either "Structural developmental anomaly of heart or great vessels, unspecified" (LA8Z), or "Other specified structural developmental anomaly of heart or great vessels" (LA8Y). Several non-highlighted terms in Table 3, such as "Bifid cardiac apex", have been crossmapped to LA8Y, when no appropriate higher order term exists in MMS.

Table 1. IPCCC ICD-11 diagnostic hierarchy

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Level 0	ICD-11 Level I	ICD-11 Level II	ICD-11 Level III	ICD-11 Level IV	ICD-11 Level V	ICD-11 Level VI	ICD-11 Level VII
1	1	01.01.59	Structural developmental anomaly of heart or great vessels							
2	2	03.01.13		Congenital anomaly of position or spatial relationships of thoraco-abdominal organs						

Table 1. (*Continued*)

3	3	02.01.09	Anomalous position-orientation of heart	
4	4**	02.01.03		Laevocardia
5	5	02.01.02		Dextrocardia
6	6	02.01.04		Mesocardia
7	7	02.01.01		Extrathoracic heart
8	8**	01.03.00	Usual atrial arrangement	
9	9	01.03.06	Abnormal atrial arrangement	
10	10	01.03.01		Atrial situs inversus
11	11	01.03.02		Isomerism of right atrial appendages
12	12	01.03.03		Isomerism of left atrial appendages
13	13	02.04.12	Abnormal ventricular relationships	
14	14**	02.03.01		Right hand pattern ventricular topology
15	15	02.03.02		Left hand pattern ventricular topology
16	16	02.03.03		Crisscross heart
17	17	02.04.00		Superior-inferior ventricular relationship
18	18	02.06.12	Abnormal relationship of great arterial roots	
19	19	02.06.03		Aortic root directly anterior to pulmonary root

(*Continued*)

Table 1. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Level 0	ICD-11 Level I	ICD-11 Level II	ICD-11 Level III	ICD-11 Level IV	ICD-11 Level V	ICD-11 Level VI	ICD-11 Level VII
20	20	02.06.02				Aortic root anterior and rightward to pulmonary root				
21	21	02.06.04				Aortic root anterior and leftward to pulmonary root				
22	22	02.06.01				Aortic root side by side and directly rightward to pulmonary root				
23	23	02.06.05				Aortic root side by side and directly leftward to pulmonary root				
24	24	02.06.07				Aortic root directly posterior to pulmonary root				
25	25**	02.06.00				Aortic root posterior and rightward to pulmonary root				
26	26	02.06.06				Aortic root posterior and leftward to pulmonary root				
27	27	02.07.03			Abnormal intrapericardial course of great arteries					
28	28**	02.07.00				Spiralling course of great arteries				
29	29	02.07.01				Parallel course of great arteries				
30	30	03.01.02			Visceral heterotaxy					
31	31	03.01.04				Right isomerism				

Table 1. (*Continued*)

32	32	03.01.05			Left Isomerism
33	33	03.01.03		Total mirror imagery	
34	34	01.03.09	Congenital anomaly of an atrioventricular or ventriculo-arterial connection		
35	35**	01.04.00		Concordant atrioventricular connections	
36	36	01.04.01		Discordant atrioventricular connections	
37	37	01.01.03			Congenitally corrected transposition of great arteries
38	38	01.05.01		Transposition of the great arteries	
39	39	01.01.02			Transposition of the great arteries with concordant atrioventricular connections and intact ventricular septum
40	40	01.01.10			Transposition of the great arteries with concordant atrioventricular connections and ventricular septal defect
41	41	01.01.10 + 07.09.01			Transposition of the great arteries with concordant atrioventricular connections and ventricular septal defect and left ventricular outflow tract obstruction

(*Continued*)

Table 1. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Level 0	ICD-11 Level I	ICD-11 Level II	ICD-11 Level III	ICD-11 Level IV	ICD-11 Level V	ICD-11 Level VI	ICD-11 Level VII
42	42**	01.05.00			Concordant ventriculo-arterial connections					
43	43	01.05.10				Concordant ventriculo-arterial connections with parallel great arteries				
44	44	01.01.04			Double outlet right ventricle					
45	45	01.01.17				Double outlet right ventricle with subaortic or doubly committed ventricular septal defect and pulmonary stenosis, Fallot type				
46	46	01.01.17 + 07.13.04					Double outlet right ventricle with subaortic ventricular septal defect and pulmonary stenosis, Fallot type			
47	47	01.01.17 + 07.13.02					Double outlet right ventricle with doubly committed ventricular septal defect and pulmonary stenosis, Fallot type			
48	48	01.01.18				Double outlet right ventricle with subpulmonary ventricular septal defect, transposition type				
49	49	01.01.19				Double outlet right ventricle with non-committed ventricular septal defect				

Table 1. (*Continued*)

50	50	01.01.40		Double outlet right ventricle with subaortic or doubly committed ventricular septal defect without pulmonary stenosis, ventricular septal defect type	
51	51	01.01.40 + 07.13.04			Double outlet right ventricle with subaortic ventricular septal defect without pulmonary stenosis
52	52	01.01.40 + 07.13.02			Double outlet right ventricle with doubly committed ventricular septal defect without pulmonary stenosis
53	53	01.01.24		Double outlet right ventricle with intact ventricular septum	
54	54	01.05.03	Double outlet left ventricle		
55	55	09.01.01	Common arterial trunk		
56	56	09.01.15		Common arterial trunk with aortic dominance	
57	57	09.01.14			Common arterial trunk with aortic dominance and both pulmonary arteries arising from trunk

(*Continued*)

Table 1. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Level 0	ICD-11 Level I	ICD-11 Level II	ICD-11 Level III	ICD-11 Level IV	ICD-11 Level V	ICD-11 Level VI	ICD-11 Level VII
58	58	09.01.11					Common arterial trunk with aortic dominance and one pulmonary artery absent from the trunk, isolated pulmonary artery			
59	59	09.01.12				Common arterial trunk with pulmonary dominance and aortic arch obstruction				
60	60	09.01.18					Common arterial trunk with pulmonary dominance and interrupted aortic arch			
61	61	09.01.19					Common arterial trunk with pulmonary dominance and aortic coarctation			
62	A+	09.02.10				Atypical truncal valve				
63	62	09.02.19					Congenital truncal valvar regurgitation			
64	63	09.02.18					Congenital truncal valvar stenosis			
65	64	09.02.01					Dysplasia of truncal valve			
66	65	04.00.07		Congenital anomaly of mediastinal vein						
67	66	04.00.08			Congenital anomaly of mediastinal systemic vein					

Table 1. (*Continued*)

68	67	04.01.09	Congenital anomaly of superior caval vein		
69	68	04.01.05		Absent right superior caval vein	
70	69	04.01.25		Left superior caval vein	
71	70	04.01.01			Left superior caval vein to coronary sinus
72	71	04.01.02			Left superior caval vein to left-sided atrium
73	B^{+}	04.01.07		Congenital stenosis of superior caval vein	
74	72	04.03.08	Congenital anomaly of inferior caval vein		
75	73	04.03.10		Interrupted inferior caval vein with absent suprarenal segment and azygos continuation	
76	C^{+}	04.03.06		Congenital stenosis of inferior caval vein	
77	74	04.04.05	Congenital anomaly of the coronary sinus		
78	75	04.04.13		Unroofed coronary sinus	
79	D^{+}	04.04.02			Completely unroofed coronary sinus
80	E^{+}	04.04.01			Partially unroofed coronary sinus
81	76	04.04.14		Coronary sinus orifice atresia or stenosis	

(*Continued*)

Table 1. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Level 0	ICD-11 Level I	ICD-11 Level II	ICD-11 Level III	ICD-11 Level IV	ICD-11 Level V	ICD-11 Level VI	ICD-11 Level VII
82	77***	04.02.13				Anomalous hepatic venous connection to heart				
83	78	04.08.04			Congenital anomaly of pulmonary vein					
84	79	04.08.07				Anomalous pulmonary venous connection				
85	80	04.08.05					Total anomalous pulmonary venous connection			
86	81	04.06.00						Total anomalous pulmonary venous connection of the supracardiac type		
87	82	04.08.10						Total anomalous pulmonary venous connection of the cardiac type		
88	83	04.08.20						Total anomalous pulmonary venous connection of the infracardiac type		
89	84	04.08.30						Total anomalous pulmonary venous connection of the mixed type		
90	85	04.07.01					Partial anomalous pulmonary venous connection			
91	86	01.01.16					Partial anomalous pulmonary venous connection of Scimitar type			

Table 1. (*Continued*)

92	87	03.02.23					Scimitar syndrome
93	88	04.08.06				Obstructed anomalous pulmonary venous pathway or connection	
94	89	04.08.31			Congenital pulmonary venous stenosis or hypoplasia		
95	90	04.08.02			Congenital atresia of pulmonary vein		
96	91	05.00.02	Congenital anomaly of an atrium or atrial septum				
97	92	05.07.01		Congenital anomaly of atrial septum			
98	93	05.06.04			Restrictive interatrial communication or intact atrial septum when an interatrial shunt is physiologically necessary		
99	94	05.03.03			Aneurysm of atrial septum		
100	95	05.04.01			Interatrial communication		
101	96**	05.03.01				Patent oval foramen	
102	97	05.04.02				Atrial septal defect within oval fossa	
103	98	05.05.00				Sinus venosus defect	
104	99	05.06.02				Common atrium with separate atrioventricular junctions	

(*Continued*)

Table 1. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Level 0	ICD-11 Level I	ICD-11 Level II	ICD-11 Level III	ICD-11 Level IV	ICD-11 Level V	ICD-11 Level VI	ICD-11 Level VII
105	100	05.05.03					Interatrial communication through coronary sinus orifice			
106	101	05.01.13			Congenital anomaly of right atrium					
107	102	05.01.21				Divided right atrium				
108	F+	05.01.04				Chiari network				
109	103	05.01.06				Left-sided juxtaposition of the atrial appendages				
110	104	05.01.12				Congenital giant right atrium				
111	105	05.02.11			Congenital anomaly of left atrium					
112	106	05.02.01				Divided left atrium				
113	107	05.02.04				Right-sided juxtaposition of the atrial appendages				
114	108	06.00.15		Congenital anomaly of an atrioventricular valve or atrioventricular septum						
115	109	06.01.11			Congenital anomaly of tricuspid valve					
116	110	06.01.25				Congenital tricuspid regurgitation				
117	111	06.01.07				Congenital tricuspid valvar stenosis				

Table 1. (*Continued*)

118	112	06.01.04		Tricuspid annular hypoplasia	
119	113	06.01.03		Dysplasia of tricuspid valve	
120	114	06.01.09		Straddling tricuspid valve	
121	115	06.01.05		Overriding tricuspid valve	
122	116	06.01.34		Ebstein malformation of tricuspid valve	
123	G^{+}	06.01.32		Absent tricuspid valve leaflet	
124	H^{+}	06.01.36		True cleft of tricuspid valve leaflet	
125	117	06.02.11	Congenital anomaly of mitral valve		
126	118	06.02.25		Congenital mitral regurgitation	
127	119	06.02.07		Congenital mitral valvar stenosis	
128	120	06.02.04		Mitral annular hypoplasia	
129	121	06.02.09		Straddling mitral valve	
130	122	06.02.05		Overriding mitral valve	
131	123	06.02.03		Dysplasia of mitral valve	
132	124	05.02.02		Supravalvar or intravalvar mitral ring	
133	I^{+}	06.02.23			Congenital intravalvar mitral ring

(*Continued*)

Table 1. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Level 0	ICD-11 Level I	ICD-11 Level II	ICD-11 Level III	ICD-11 Level IV	ICD-11 Level V	ICD-11 Level VI	ICD-11 Level VII
134	J+	06.02.17					Congenital supravalvar mitral ring			
135	125	06.02.72				Congenital mitral valvar prolapse				
136	126	06.02.36				True cleft of anterior mitral leaflet				
137	127	06.02.21				Congenital anomaly of mitral subvalvar apparatus				
138	128	06.02.22					Congenital mitral subvalvar stenosis			
139	129	06.02.56					Parachute malformation of mitral valve			
140	K+	06.02.39				Accessory tissue on mitral valve leaflet				
141	L+	06.02.32				Congenital unguarded mitral orifice				
142	M+	06.02.33				Double orifice of mitral valve				
143	130	06.04.11			Congenital anomaly of left-sided atrioventricular valve in double inlet ventricle					
144	131	06.03.11			Congenital anomaly of right-sided atrioventricular valve in double inlet ventricle					
145	132	06.06.11			Common atrioventricular junction					

Table 1. (*Continued*)

146	133	06.06.00	Common atrioventricular junction with atrioventricular septal defect		
147	134	06.07.27		Atrioventricular septal defect with balanced ventricles	
148	135	06.07.26		Atrioventricular septal defect with ventricular imbalance	
149	136	06.07.05			Atrioventricular septal defect with ventricular imbalance with dominant right ventricle and hypoplastic left ventricle
150	137	06.07.06			Atrioventricular septal defect with ventricular imbalance with dominant left ventricle and hypoplastic right ventricle
151	138	06.06.01		Atrioventricular septal defect with communication at the atrial level only	
152	139	06.06.08		Atrioventricular septal defect with communication at the ventricular level only	
153	140	06.06.10		Atrioventricular septal defect with communication at atrial level and restrictive communication at ventricular level	

(*Continued*)

Table 1. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Level 0	ICD-11 Level I	ICD-11 Level II	ICD-11 Level III	ICD-11 Level IV	ICD-11 Level V	ICD-11 Level VI	ICD-11 Level VII
154	141	06.06.09					Atrioventricular septal defect with communication at atrial level and unrestrictive communication at ventricular level			
155	142	01.01.20					Atrioventricular septal defect and tetralogy of Fallot			
156	143	05.06.03					Common atrium with common atrioventricular junction			
157	144	06.05.60					Common atrioventricular valvar regurgitation			
158	145	06.05.14					Atypical common atrioventricular valve			
159	146	06.07.36						Common atrioventricular valve with unbalanced commitment of valve to ventricles		
160	147	06.07.37							Common atrioventricular valve with unbalanced commitment of valve to right ventricle	
161	148	06.07.38							Common atrioventricular valve with unbalanced commitment of valve to left ventricle	

Table 1. (*Continued*)

162	149	06.05.71					Atypical right ventricular component of common atrioventricular valve	
163	150	06.05.72					Atypical left ventricular component of common atrioventricular valve	
164	151	06.05.25						Double orifice of left ventricular component of common atrioventricular valve
165	152	06.05.98						Deficient mural leaflet of left ventricular component of common atrioventricular valve
166	153	06.07.28			Common atrioventricular junction without an atrioventricular septal defect			
167	154	07.14.02		Communication between left ventricle and right atrium				
168	155	07.00.00	Congenital anomaly of a ventricle or the ventricular septum					
169	156	07.01.07		Congenital right ventricular anomaly				
170	157	07.02.00			Right ventricular hypoplasia			

(*Continued*)

Table 1. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Level 0	ICD-11 Level I	ICD-11 Level II	ICD-11 Level III	ICD-11 Level IV	ICD-11 Level V	ICD-11 Level VI	ICD-11 Level VII
171	158	07.05.20				Congenital right ventricular outflow tract obstruction				
172	159	07.03.01				Double chambered right ventricle				
173	160	07.01.13				Right ventricular myocardial sinusoids				
174	161	07.01.06				Parchment right ventricle				
175	162	01.01.01				Tetralogy of Fallot				
176	163	09.05.25					Tetralogy of Fallot with absent pulmonary valve syndrome			
177	164	01.01.26					Tetralogy of Fallot with pulmonary atresia			
178	165	01.01.57					Tetralogy of Fallot with pulmonary atresia and systemic-to-pulmonary collateral arteries			
179	166	07.06.07			Congenital left ventricular anomaly					
180	167	07.07.00				Left ventricular hypoplasia				
181	168	07.06.19				Congenital left ventricular aneurysm or diverticulum				
182	N^{+}	07.06.01					Congenital left ventricular aneurysm			

Table 1. (*Continued*)

183	O^+	07.06.03			Congenital left ventricular diverticulum
184	169	07.09.28		Congenital left ventricular outflow tract obstruction	
185	170	07.09.08			Congenital left ventricular outflow tract obstruction due to atrioventricular valve
186	171	01.01.33		Left heart obstruction at multiple sites	
187	172	07.06.12		Left ventricular myocardial sinusoids	
188	P^+	07.00.07	Anomalous ventricular bands		
189	173	07.20.04	Congenital anomaly of ventricular septum		
190	174	07.14.07		Restrictive interventricular communication when an interventricular shunt is physiologically necessary	
191	175	07.10.00		Ventricular septal defect	
192	176	07.10.01			Perimembranous central ventricular septal defect
193	177	07.14.05			Inlet ventricular septal defect without a common atrioventricular junction

(*Continued*)

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Table 1. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Level 0	ICD-11 Level I	ICD-11 Level II	ICD-11 Level III	ICD-11 Level IV	ICD-11 Level V	ICD-11 Level VI	ICD-11 Level VII
194	178	07.10.02						Inlet perimembranous ventricular septal defect without atrioventricular septal malalignment without a common atrioventricular junction		
195	179	07.14.06						Inlet perimembranous ventricular septal defect with atrioventricular septal malalignment and without a common atrioventricular junction		
196	180	07.11.02						Inlet muscular ventricular septal defect		
197	181	07.11.01					Trabecular muscular ventricular septal defect			
198	182	07.11.04						Trabecular muscular ventricular septal defect midseptal		
199	183	07.11.03						Trabecular muscular ventricular septal defect apical		
200	184	07.11.12						Trabecular muscular ventricular septal defect postero-inferior		

Table 1. (*Continued*)

201	185	07.11.07			Trabecular muscular ventricular septal defect anterosuperior		
202	186	07.11.05			Multiple trabecular muscular ventricular septal defects		
203	187	07.12.00		Outlet ventricular septal defect			
204	188	07.12.09			Outlet ventricular septal defect without malalignment		
205	189	07.11.06				Outlet muscular ventricular septal defect without malalignment	
206	190	07.12.01				Doubly committed juxta-arterial ventricular septal defect without malalignment	
207	191	07.12.02					Doubly committed juxta-arterial ventricular septal defect without malalignment and with muscular postero-inferior rim
208	192	07.12.03					Doubly committed juxta-arterial ventricular septal defect without malalignment and with perimembranous extension
209	193	07.10.17			Outlet ventricular septal defect with anteriorly malaligned outlet septum		

(*Continued*)

Table 1. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Level 0	ICD-11 Level I	ICD-11 Level II	ICD-11 Level III	ICD-11 Level IV	ICD-11 Level V	ICD-11 Level VI	ICD-11 Level VII
210	194	07.11.15							Outlet muscular ventricular septal defect with anteriorly malaligned outlet septum	
211	195	07.10.04							Outlet perimembranous ventricular septal defect with anteriorly malaligned outlet septum	
212	196	07.12.12							Doubly committed juxta-arterial ventricular septal defect with anteriorly malaligned fibrous outlet septum	
213	197	07.12.07								Doubly committed juxta-arterial ventricular septal defect with anteriorly malaligned fibrous outlet septum and muscular postero-inferior rim
214	198	07.12.05								Doubly committed juxta-arterial ventricular septal defect with anteriorly malaligned fibrous outlet septum and perimembranous extension
215	199	07.10.18						Outlet ventricular septal defect with posteriorly malaligned outlet septum		

Table 1. (*Continued*)

216	200	07.11.16			Outlet muscular ventricular septal defect with posteriorly malaligned outlet septum	
217	201	07.10.19			Outlet perimembranous ventricular septal defect with posteriorly malaligned outlet septum	
218	202	07.12.13			Doubly committed juxta-arterial ventricular septal defect with posteriorly malaligned fibrous outlet septum	
219	203	07.12.08				Doubly committed juxta-arterial ventricular septal defect with posteriorly malaligned fibrous outlet septum and muscular postero-inferior rim
220	204	07.12.06				Doubly committed juxta-arterial ventricular septal defect with posteriorly malaligned fibrous outlet septum and perimembranous extension
221	205	07.15.01		Ventricular septal defect haemodynamically insignificant		

(*Continued*)

Table 1. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Level 0	ICD-11 Level I	ICD-11 Level II	ICD-11 Level III	ICD-11 Level IV	ICD-11 Level V	ICD-11 Level VI	ICD-11 Level VII
222	206	07.15.04					Multiple ventricular septal defects			
223	207	01.01.22		Functionally univentricular heart						
224	208	01.01.14			Double inlet atrioventricular connection					
225	209	01.04.04				Double inlet left ventricle				
226	210	01.04.03				Double inlet right ventricle				
227	211	01.04.05				Double inlet to solitary ventricle of indeterminate morphology				
228	212	06.01.01			Tricuspid atresia					
229	213	06.01.26				Tricuspid atresia with absent atrioventricular connection				
230	214	06.01.02				Tricuspid atresia with imperforate tricuspid valve				
231	215	06.02.01			Mitral atresia					
232	216	06.02.26				Mitral atresia with absent atrioventricular connection				
233	217	06.02.02				Mitral atresia with imperforate mitral valve				
234	218	01.01.09			Hypoplastic left heart syndrome					
235	219	09.04.29		Congenital anomaly of a ventriculo-arterial valve or adjacent regions						

Table 1. (*Continued*)

236	220	09.05.29	Congenital anomaly of pulmonary valve	
237	221	09.05.04		Congenital pulmonary valvar stenosis
238	222	09.05.05		Pulmonary annular hypoplasia
239	223	09.05.22		Congenital pulmonary regurgitation
240	224	09.05.24		Dysplasia of pulmonary valve
241	225	09.05.32		Bicuspid pulmonary valve
242	226	07.05.32	Congenital subpulmonary stenosis	
243	227	09.07.15	Congenital supravalvar pulmonary stenosis	
244	228	09.05.16	Congenital pulmonary atresia	
245	Q+	09.05.12		Congenital pulmonary valvar atresia
246	229	01.01.07		Pulmonary atresia with intact ventricular septum
247	230	09.15.19	Congenital anomaly of aortic valve	
248	231	09.15.01		Congenital aortic valvar stenosis
249	232	09.15.07		Congenital aortic regurgitation
250	233	09.15.22		Bicuspid aortic valve
251	234	09.15.21		Unicuspid aortic valve

(*Continued*)

Table 1. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Level 0	ICD-11 Level I	ICD-11 Level II	ICD-11 Level III	ICD-11 Level IV	ICD-11 Level V	ICD-11 Level VI	ICD-11 Level VII
252	235***	09.15.30				Aortic valvar prolapse				
253	236	09.15.06				Aortic valvar atresia				
254	237	09.15.17				Aortic annular hypoplasia				
255	238	09.15.09				Dysplasia of aortic valve				
256	239	07.09.50			Congenital subaortic stenosis					
257	240	07.09.03				Subaortic stenosis due to fibromuscular shelf				
258	241	07.09.16				Subaortic stenosis due to fibromuscular tunnel				
259	242	09.16.18			Congenital supravalvar aortic stenosis					
260	243	09.18.01			Aneurysm of aortic sinus of Valsalva					
261	244	09.17.01			Aortoventricular tunnel					
262	R^+	09.17.02				Aorto-left ventricular tunnel				
263	S^+	09.17.04				Aorto-right ventricular tunnel				
264	245	09.04.28		Congenital anomaly of great arteries including arterial duct						

Table 1. (*Continued*)

265	246	09.04.07	Congenital aortopulmonary window			
266	247	09.07.16	Congenital anomaly of pulmonary arterial tree			
267	248	09.10.36		Congenital dilation of pulmonary arterial tree		
268	249	09.07.19		Congenital pulmonary trunk anomaly		
269	250	09.07.20			Congenital pulmonary trunk hypoplasia	
270	251	09.07.05			Absent or atretic pulmonary trunk	
271	252	09.10.41		Congenital pulmonary arterial branch anomaly		
272	253	09.10.27			Congenital pulmonary arterial branch stenosis	
273	254	09.10.28				Congenital right pulmonary arterial stenosis
274	255	09.10.29				Congenital left pulmonary arterial stenosis
275	256	09.10.71			Congenital pulmonary arterial branch hypoplasia	
276	257	09.10.72				Congenital right pulmonary arterial hypoplasia
277	258	09.10.73				Congenital left pulmonary arterial hypoplasia

(*Continued*)

Table 1. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Level 0	ICD-11 Level I	ICD-11 Level II	ICD-11 Level III	ICD-11 Level IV	ICD-11 Level V	ICD-11 Level VI	ICD-11 Level VII
278	T+	09.10.21					Absent or atretic right or left pulmonary artery			
279	259	09.10.75						Absent or atretic right pulmonary artery		
280	260	09.10.77						Absent or atretic left pulmonary artery		
281	261	09.10.37					Congenital central pulmonary arterial stenosis or hypoplasia proximal to hilar bifurcation			
282	262	09.10.38					Congenital peripheral pulmonary arterial stenosis or hypoplasia at or beyond hilar bifurcation			
283	263	09.10.30					Congenitally discontinuous, non-confluent right and left pulmonary arteries			
284	264	09.09.08					Pulmonary artery origin from ascending aorta			
285	265	09.09.03						Right pulmonary artery from ascending aorta		
286	266	09.09.05						Left pulmonary artery from ascending aorta		
287	267	09.09.11					Pulmonary artery from arterial duct			
288	268	09.09.02						Right pulmonary artery from arterial duct		

Table 1. (*Continued*)

289	269	09.09.04				Left pulmonary artery from arterial duct
290	270	07.09.34	Congenital anomaly of aorta or its branches			
291	271	09.16.06		Congenital anomaly of ascending aorta		
292	272	09.16.02			Hypoplasia of ascending aortic	
293	273	09.16.19			Congenital ascending aortic aneurysm or dilation	
294	274	09.28.10		Congenital anomaly of aortic arch		
295	275	09.29.11			Hypoplasia of aortic arch	
296	277	09.29.31			Interrupted aortic arch	
297	278	09.29.32				Interrupted aortic arch distal to subclavian artery, type A
298	279	09.29.33				Interrupted aortic arch between subclavian and common carotid arteries, type B
299	280	09.29.34				Interrupted aortic arch between carotid arteries, type C
300	281	09.28.15			Right aortic arch	
301	282	09.28.22			Left aortic arch	
302	283	09.28.06			Cervical aortic arch	
303	X^+	09.30.22			Aortic diverticulum of Kommerell	
304	Y^+	09.28.08			Persistent fifth aortic arch	

(*Continued*)

Table 1. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Level 0	ICD-11 Level I	ICD-11 Level II	ICD-11 Level III	ICD-11 Level IV	ICD-11 Level V	ICD-11 Level VI	ICD-11 Level VII
305	276	09.29.01				Coarctation of aorta				
306	U+	09.29.02					Preductal coarctation of aorta			
307	V+	09.29.04					Postductal coarctation of aorta			
308	W+	09.29.03					Juxtaductal (paraductal) coarctation of aorta			
309	284	09.30.17				Congenital anomaly of aortic arch branch				
310	285	09.30.02					Aberrant origin of right subclavian artery			
311	286	09.30.04					Aberrant origin of left subclavian artery			
312	287	09.30.16					Isolation of an aortic arch branch			
313	Z+	09.30.11						Isolation of innominate artery		
314	AA+	09.30.14						Isolation of left subclavian artery		
315	AB+	09.30.15						Isolation of right subclavian artery		
316	AC+	09.30.12						Isolation of left common carotid artery		
317	AD+	09.30.13						Isolation of right common carotid artery		
318	AE+	09.30.34					Aberrant origin of innominate artery			

Table 1. (*Continued*)

319	AF+	09.30.31			Common origin of the innominate artery and left common carotid artery	
320	AG+	09.30.28			Separate origins of internal and external carotid arteries	
321	288	09.28.47		Congenital anomaly of descending thoracic or abdominal aorta		
322	289	09.29.44			Descending thoracic or abdominal aortic coarctation	
323	AH+	09.29.05				Coarctation of the descending thoracic aorta
324	AI+	09.29.06				Coarctation of the abdominal aorta
325	290	09.31.40	Tracheo-oesophageal compressive syndrome			
326	291	09.30.23		Innominate artery compression syndrome		
327	AJ+	09.30.27			Retro-oesophageal origin of aberrant innominate artery	
328	292	09.31.00	Vascular Ring			
329	293	09.28.09		Double aortic arch		
330	294	09.31.35		Vascular ring of right aortic arch and left arterial duct or ligament		
331	295	09.31.34		Vascular ring of left aortic arch and right arterial duct or ligament		

(*Continued*)

Table 1. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Level 0	ICD-11 Level I	ICD-11 Level II	ICD-11 Level III	ICD-11 Level IV	ICD-11 Level V	ICD-11 Level VI	ICD-11 Level VII
332	296	09.09.06			Anomalous origin of left pulmonary artery from right pulmonary artery					
333	297	09.27.05			Congenital arterial duct anomaly					
334	298	09.27.21				Patent arterial duct				
335	AK^{+}	09.27.03				Absent arterial duct				
336	AL^{+}	09.27.04				Congenital aneurysm of arterial duct				
337	AM^{+}	09.27.41				Anomalous origin of arterial duct				
338	AN^{+}	09.27.82				Anomalous origin of arterial ligament				
339	AO^{+}	14.10.51				Fetal arterial duct narrowing-closure				
340	299	09.08.18			Systemic-to-pulmonary collateral arteries					
341	300	09.46.03		Congenital anomaly of coronary artery						
342	301	09.41.01			Anomalous origin of coronary artery from pulmonary arterial tree					
343	302	09.41.03				Anomalous origin of left coronary artery from pulmonary artery				

Table 1. (*Continued*)

344	303	09.42.00	Anomalous aortic origin or course of coronary artery		
345	304	09.42.21		Anomalous aortic origin of coronary artery with ventriculo-arterial concordance	
346	305	09.46.26			Right coronary artery from left aortic sinus with ventriculo-arterial concordance
347	306	09.46.21			Left coronary artery from right aortic sinus with ventriculo-arterial concordance
348	307	09.43.04		Anterior descending from right coronary artery across right ventricular outflow tract	
349	308	09.43.05		Intramural proximal coronary arterial course	
350	309	09.43.13		Single coronary artery supplying all of heart	
351	310	09.43.12	Myocardial bridging of coronary artery		
352	311	09.44.05	Congenital coronary arterial orifice stenosis		
353	312	09.44.19	Congenital coronary arterial orifice atresia		
354	313	09.45.16	Congenital coronary arterial fistula		

(*Continued*)

Table 1. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Level 0	ICD-11 Level I	ICD-11 Level II	ICD-11 Level III	ICD-11 Level IV	ICD-11 Level V	ICD-11 Level VI	ICD-11 Level VII
355	314	09.45.10				Congenital coronary arterial fistula to right ventricle				
356	315	09.45.22				Congenital coronary arterial fistula to left ventricle				
357	316	09.46.14			Congenital coronary arterial aneurysm					
358	AP^{+}	09.42.09			Accessory coronary artery					
359	AQ^{+}	09.46.44			Congenital absence of coronary artery					
360	AR^{+}	09.46.19			Coronary arterial hypoplasia					
361	317	10.01.05		Congenital pericardial anomaly						
362	AS^{+}	10.01.02			Complete agenesis of pericardium					
363	AT^{+}	10.01.01			Partial agenesis of pericardium					
364	AU^{+}	10.01.03			Pleuropericardial cyst					
365	AV^{+}	10.03.53		Congenital cardiac tumour						
366	318***	09.19.05		Pulmonary arteriovenous fistula						
367	AW^{+}	02.02.03		Bifid apex of heart						

Table 2. IPCCC ICD-11 Definitions

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
1	1	01.01.59	Structural developmental anomaly of heart or great vessels	A congenital malformation of the heart and/or great vessels or an acquired abnormality unique to the congenitally malformed heart.	This term should be selected only if a more specific term does not exist.	Congenital anomaly of heart and/or great vessels and related acquired abnormality; Congenital heart disease; Congenital malformation of heart	
2	2	03.01.13	Congenital anomaly of position or spatial relationships of thoraco-abdominal organs	A congenital cardiovascular finding or malformation associated with an abnormal position of the heart or thoraco-abdominal organs, or an abnormal relative position of its component parts.			
3	3	02.01.09	Anomalous position-orientation of heart	A congenital cardiovascular finding/malformation in which there is an abnormality of the position or orientation of heart.		Malposition of heart	
4	4**	02.01.03	Laevocardia	A congenital cardiovascular finding in which the heart is predominantly to the left of the thoracic midline.	This is independent of the orientation of the cardiac apex. This is a normal finding and should be coded only in the context of complex heart disease.	Levocardia; Left-sided heart	
5	5	02.01.02	Dextrocardia	A congenital cardiovascular malformation in which the heart is predominantly to the right of the thoracic midline.	This is independent of the orientation of the cardiac apex.	Right-sided heart; heart in right chest; congenital dextrocardia of heart	
6	6	02.01.04	Mesocardia	A congenital cardiovascular malformation in which the heart is central or midline within the thorax.		Midline heart	
7	7	02.01.01	Extrathoracic heart	A congenital cardiovascular malformation in which the heart is at least partially outside of the thorax.		Ectopia cordis	
8	8**	01.03.00	Usual atrial arrangement	A congenital cardiac finding in which the atrial laterality (sidedness) is normal.	This is a normal finding and should be coded only in the context of complex congenital heart disease.	Atrial situs solitus	
9	9	01.03.06	Abnormal atrial arrangement	A congenital cardiac malformation in which there is an abnormality of the laterality (or sidedness) of the atria.		Abnormal atrial situs	
10	10	01.03.01	Atrial situs inversus	A congenital cardiac malformation in which the atrial morphologies and positions are the mirror image of normal.		Mirror-image atrial arrangement	

(*Continued*)

Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
11	11	01.03.02	Isomerism of right atrial appendages	A congenital cardiac malformation in which both atrial appendages have the morphology of a right atrial appendage.		Bilateral right atrial appendages, isomeric right atrial appendages	
12	12	01.03.03	Isomerism of left atrial appendages	A congenital cardiac malformation in which both atrial appendages have the morphology of a left atrial appendage.		Bilateral left atrial appendages, isomeric left atrial appendages	
13	13	02.04.12	Abnormal ventricular relationships	A congenital cardiac malformation in which the ventricular positions relative to each other or their laterality (sidedness) are abnormal.			
14	14**	02.03.01	Right hand pattern ventricular topology	A congenital cardiac finding in which the chirality, or handedness, of the ventricles is normal, also known as D-loop.	Chirality or handedness with a right-handed right ventricle and means likening the morphological right ventricle to a right hand by representing the inflow by the thumb, outflow by the index finger, and septum by the palm. This is a normal finding but should be coded in the presence of abnormal positions of the atria or great arteries where it represents an abnormality of the interrelationships of the ventricles relative to the remainder of the heart.	D-loop ventricles, Dextro-ventricular looping, D-bulboventricular loop	
15	15	02.03.02	Left hand pattern ventricular topology	A congenital cardiac malformation in which the chirality, or handedness, of the ventricles is mirror image of normal, also known as L-loop.	Chirality or handedness with a left-handed right ventricle and means likening the morphological right ventricle to a left hand by representing the inflow by the thumb, outflow by the index finger, and septum by the palm. An example is congenitally corrected transposition of great arteries.	L-loop ventricles, Levo-ventricular looping, Sinistro-ventricular looping, L-bulboventricular loop	
16	16	02.03.03	Crisscross heart	A congenital cardiac malformation in which the atrioventricular inflow vectors are approximately orthogonal or perpendicular.		Twisted atrioventricular connections; Criss-cross heart	
17	17	02.04.00	Superior-inferior ventricular relationship	A congenital cardiac malformation in which the ventricles are positioned superior-inferior to each other.	Excludes situations where one ventricle does not receive an atrioventricular valve (univentricular atrioventricular connection).	Upstairs-downstairs ventricular relationship, supero-inferior heart	

Table 2. (*Continued*)

18	18	02.06.12	Abnormal relationship of great arterial roots	A congenital cardiovascular malformation in which the aortic root or its remnant is abnormally positioned relative to the pulmonary root or its remnant.		Abnormal relationships of great arteries, Abnormal relationships of great vessels
19	19	02.06.03	Aortic root directly anterior to pulmonary root	A congenital cardiovascular malformation in which the aortic root or its remnant is positioned directly anterior to the pulmonary root or its remnant.		Antero-posterior great arteries, Antero-posterior great vessels, A-malposed aorta, A-malposed great arteries, Aorta directly anterior to pulmonary artery
20	20	02.06.02	Aortic root anterior and rightward to pulmonary root	A congenital cardiovascular malformation in which the aortic root or its remnant is positioned anterior and to the right of the pulmonary root or its remnant.		D-malposed great arteries with anterior aorta, D-malposed great vessels with anterior aorta, Dextroposed great arteries with anterior aorta, Dextroposed great vessels with anterior aorta, D-transposed great arteries, D-transposed great vessels
21	21	02.06.04	Aortic root anterior and leftward to pulmonary root	A congenital cardiovascular malformation in which the aortic root or its remnant is positioned anterior and to the left of the pulmonary root or its remnant.		L-malposed great arteries with anterior aorta, L-malposed great vessels with anterior aorta, Levoposed great arteries with anterior aorta, Levoposed great vessels with anterior aorta, L-transposed great arteries with anterior aorta, L-transposed great vessels with anterior aorta
22	22	02.06.01	Aortic root side by side and directly rightward to pulmonary root	A congenital cardiovascular malformation in which the aortic root or its remnant is positioned directly to the right of the pulmonary root or its remnant.		D-malposed side-by-side great arteries, D-malposed side-by-side great vessels, Dextroposed side-by-side great arteries, Dextroposed side-by-side great vessels
23	23	02.06.05	Aortic root side by side and directly leftward to pulmonary root	A congenital cardiovascular malformation in which the aortic root or its remnant is positioned directly to the left of the pulmonary root or its remnant.		L-malposed side-by-side great arteries, L-malposed side-by-side great vessels, Levoposed side-by-side great arteries, Levoposed side-by-side great vessels
24	24	02.06.07	Aortic root directly posterior to pulmonary root	A congenital cardiovascular malformation in which the aortic root or its remnant is positioned directly posterior to the pulmonary root or its remnant.		
25	25**	02.06.00	Aortic root posterior and rightward to pulmonary root	A congenital cardiovascular finding in which the aortic root or its remnant is positioned posterior and to the right of the pulmonary root or its remnant.	This is a normal finding but should only be coded in the context of complex congenital heart disease.	Segmental analysis D with posterior aorta, Normally related great arteries, Normally related great vessels, D-malposed great arteries with posterior aorta, D-malposed great vessels with posterior aorta

(*Continued*)

Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
26	26	02.06.06	Aortic root posterior and leftward to pulmonary root	A congenital cardiovascular malformation in which the aortic root or its remnant is positioned posterior and to the left of the pulmonary root or its remnant.		L-malposed great arteries with posterior aorta, L-malposed great vessels with posterior aorta, Levoposed great arteries, Levoposed great vessels, Mirror imaged normally related great arteries, Mirror imaged normally related great vessels	
27	27	02.07.03	Abnormal intrapericardial course of great arteries	A congenital cardiovascular malformation in which the course of the ascending aorta or its remnant is abnormal relative to the course of the pulmonary trunk or its remnant.	The normal course of the proximal great vessels results in a nearly orthogonal relationship of the axis of the aorta to the axis of the pulmonary artery.		
28	28**	02.07.00	Spiralling course of great arteries	A congenital cardiovascular finding in which the course of the great arteries results in a nearly orthogonal relationship of the axis of the ascending aorta to the axis of the pulmonary trunk.	This is a normal finding but should be coded in the presence of abnormal positions of the great arteries relative to the ventricles.	Normal course of great arteries	
29	29	02.07.01	Parallel course of great arteries	A congenital cardiovascular finding in which the course of the great arteries results in a nearly parallel relationship of the axis of the ascending aorta to the axis of the pulmonary trunk.			
30	30	03.01.02	Visceral heterotaxy	A congenital malformation in which the internal thoraco-abdominal organs demonstrate abnormal arrangement across the left-right axis of the body. By convention, in congenital cardiology, heterotaxy syndrome does not include patients with complete mirror-imaged arrangement of the internal organs along the left-right axis also known as “total mirror imagery” or “situs inversus totalis”.		Situs ambiguus; Situs ambiguous; Heterotaxy; Heterotaxy syndrome, Heterotaxia; Heterotaxia syndrome; Splenic syndromes; Abnormal arrangement of thoraco-abdominal organs	
31	31	03.01.04	Right isomerism	A congenital cardiovascular malformation that is a variant of heterotaxy syndrome in which some paired structures on opposite sides of the left-right axis of the body are symmetrical mirror images of each other and have the morphology of the normal right-sided structures.		Bilateral right-sidedness; Ivemark syndrome; Asplenia syndrome; Splenic agenesis syndrome	

Table 2. (*Continued*)

32	32	03.01.05	Left Isomerism	A congenital cardiovascular malformation that is a variant of an heterotaxy syndrome in which some paired structures on opposite sides of the left-right axis of the body are symmetrical mirror images of each other and have the morphology of the normal left-sided structures.		Bilateral left-sidedness; Polysplenia syndrome; Moller syndrome	
33	33	03.01.03	Total mirror imagery	A congenital malformation in which there is complete mirror-imaged arrangement of the internal organs along the left-right axis of the body.		Situs inversus totalis; Situs inversus viscerum; Situs inversus	
34	34	01.03.09	Congenital anomaly of an atrioventricular or ventriculo-arterial connection	A congenital cardiovascular malformation in which one or more of the following connections is abnormal: 1) the morphologically right atrium to the morphologically right ventricle, 2) the morphologically left atrium to the morphologically left ventricle, 3) the morphologically right ventricle to the pulmonary trunk, 4) the morphologically left ventricle to the aorta.	This excludes codes for hearts with a univentricular atrioventricular connection (mitral atresia, tricuspid atresia and double inlet ventricle), as these are listed under Functionally Univentricular Heart.		
35	35**	01.04.00	Concordant atrioventricular connections	A congenital cardiovascular finding in which the morphologically right atrium connects to the morphologically right ventricle and the morphologically left atrium connects to the morphologically left ventricle.	This is a normal finding that should only be coded when associated with abnormal atrial arrangement and/or ventriculo-arterial connections.	Normal atrioventricular connections; Atrioventricular concordance	
36	36	01.04.01	Discordant atrioventricular connections	A congenital cardiac malformation in which the morphologically right atrium connects to the morphologically left ventricle and the morphologically left atrium connects to the morphologically right ventricle.		Atrioventricular discordance	
37	37	01.01.03	Congenitally corrected transposition of great arteries	A congenital cardiovascular malformation in which the morphologically right atrium connects to the morphologically left ventricle, the morphologically left atrium connects to the morphologically right ventricle, the morphologically right ventricle connects to the aorta, and the morphologically left ventricle connects to the pulmonary trunk.		Transposition {S,L,L}; Transposition {I,D,D}; Discordant atrioventricular & ventriculo-arterial connections; Corrected transposition of the great arteries; Corrected transposition of the great vessels; Congenitally corrected transposition of the great vessels; Double discordance; Physiologically corrected transposition of the great vessels; Physiologically corrected transposition of the great arteries	ccTGA

(*Continued*)

Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
38	38	01.05.01	Transposition of the great arteries	A congenital cardiovascular malformation in which the morphologically right ventricle connects to the aorta and the morphologically left ventricle connects to the pulmonary trunk.		Discordant ventriculo-arterial connections; Transposition of the great vessels	TGA; TGV
39	39	01.01.02	Transposition of the great arteries with concordant atrioventricular connections and intact ventricular septum	A congenital cardiovascular malformation in which the morphologically right atrium connects to the morphologically right ventricle, the morphologically left atrium connects to the morphologically left ventricle, the morphologically right ventricle connects to the aorta, the morphologically left ventricle connects to the pulmonary trunk, and a ventricular septal defect is not present.		Transposition {S,D,D}; Transposition {I,L,L}; Concordant atrioventricular and discordant ventriculo-arterial connections with intact ventricular septum; Simple transposition	TGA & IVS; D-TGA
40	40	01.01.10	Transposition of the great arteries with concordant atrioventricular connections and ventricular septal defect	A congenital cardiovascular malformation in which the morphologically right atrium connects to the morphologically right ventricle, the morphologically left atrium connects to the morphologically left ventricle, the morphologically right ventricle connects to the aorta, the morphologically left ventricle connects to the pulmonary trunk, and one or more ventricular septal defects are present.		Concordant atrioventricular connections and discordant ventriculo-arterial connections with ventricular septal defect	TGA & VSD
41	41	01.01.10 + 07.09.01	Transposition of the great arteries with concordant atrioventricular connections and ventricular septal defect and left ventricular outflow tract obstruction	A congenital cardiovascular malformation in which the morphologically right atrium connects to the morphologically right ventricle, the morphologically left atrium connects to the morphologically left ventricle, the morphologically right ventricle connects to the aorta, the morphologically left ventricle connects to the pulmonary trunk, one or more ventricular septal defects are present, and left ventricular outflow tract obstruction is present.		Concordant atrioventricular connections and discordant ventriculo-arterial connections with ventricular septal defect and left ventricular outflow tract obstruction; Transposition with ventricular septal defect and pulmonary stenosis	TGA & VSD & LVOTO; TGA & VSD & PS

Table 2. (*Continued*)

42	42**	01.05.00	Concordant ventriculo-arterial connections	A congenital cardiovascular finding in which the left ventricle is connected to the aorta or its remnant and the right ventricle is connected to the pulmonary trunk or its remnant.	This is a normal finding that should only be coded when associated with abnormal atrioventricular connections and/or ventriculo-arterial arrangement	Concordant ventriculo-arterial alignments	
43	43	01.05.10	Concordant ventriculo-arterial connections with parallel great arteries	A congenital cardiovascular malformation in which the left ventricle is connected by way of an infundibulum to the aorta or its remnant, the right ventricle is connected to the pulmonary trunk or its remnant, and the proximal arteries have a parallel course.		Anatomically corrected malposition of the great arteries	ACM
44	44	01.01.04	Double outlet right ventricle	A congenital cardiovascular malformation in which both great arteries arise entirely or predominantly from the morphologically right ventricle.			DORV
45	45	01.01.17	Double outlet right ventricle with subaortic or doubly committed ventricular septal defect and pulmonary stenosis, Fallot type	A congenital cardiovascular malformation that is a variant of double outlet right ventricle with concordant atrioventricular connections, a subaortic or doubly committed (with absence or deficiency of the conal septum) ventricular septal defect, and pulmonary outflow tract obstruction.		Double outlet right ventricle with subaortic or doubly committed interventricular communication & pulmonary stenosis (Fallot type)	DORV & PS
46	46	01.01.17 + 07.13.04	Double outlet right ventricle with subaortic ventricular septal defect and pulmonary stenosis, Fallot type	A congenital cardiovascular malformation that is a variant of double outlet right ventricle with concordant atrioventricular connections, a subaortic ventricular septal defect, and pulmonary outflow tract obstruction.		Double outlet right ventricle with subaortic interventricular communication & pulmonary stenosis (Fallot type)	DORV & PS
47	47	01.01.17 + 07.13.02	Double outlet right ventricle with doubly committed ventricular septal defect and pulmonary stenosis, Fallot type	A congenital cardiovascular malformation that is a variant of double outlet right ventricle with concordant atrioventricular connections, a doubly committed ventricular septal defect with absence or deficiency of the conal septum, and pulmonary outflow tract obstruction.		Double outlet right ventricle with doubly committed interventricular communication & pulmonary stenosis (Fallot type)	DORV & PS
48	48	01.01.18	Double outlet right ventricle with subpulmonary ventricular septal defect, transposition type	A congenital cardiovascular malformation that is a variant of double outlet right ventricle with concordant atrioventricular connections that is associated with a subpulmonary ventricular septal defect (includes Taussig-Bing heart).		Double outlet right ventricle with subpulmonary interventricular communication (transposition type); Taussig-Bing syndrome; Taussig-Bing malformation; Taussig-Bing heart	

(*Continued*)

Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
49	49	01.01.19	Double outlet right ventricle with non-committed ventricular septal defect	A congenital cardiovascular malformation that is a variant of double outlet right ventricle with concordant atrioventricular connections that is associated with ventricular septal defect that is remote from the ventricular outflow tracts and usually within the inlet or muscular septum.		Double outlet right ventricle with remote ventricular septal defect; Double outlet right ventricle with uncommitted ventricular septal defect; Double outlet right ventricle with non-committed interventricular communication	
50	50	01.01.40	Double outlet right ventricle with subaortic or doubly committed ventricular septal defect without pulmonary stenosis, ventricular septal defect type	A congenital cardiovascular malformation that is a variant of double outlet right ventricle with concordant atrioventricular connections, a subaortic or doubly committed (with absence or deficiency of the conal septum) ventricular septal defect, and unobstructed pulmonary outflow tract.		Double outlet right ventricle with subaortic or doubly committed interventricular communication without pulmonary stenosis (ventricular septal defect type)	
51	51	01.01.40 + 07.13.04	Double outlet right ventricle with subaortic ventricular septal defect without pulmonary stenosis	A congenital cardiovascular malformation that is a variant of double outlet right ventricle with concordant atrioventricular connections, a subaortic or doubly committed (with absence or deficiency of the conal septum) ventricular septal defect, and unobstructed pulmonary outflow tract.		Double outlet right ventricle with subaortic or doubly committed interventricular communication without pulmonary stenosis (ventricular septal defect type)	
52	52	01.01.40 + 07.13.02	Double outlet right ventricle with doubly committed ventricular septal defect without pulmonary stenosis	A congenital cardiovascular malformation that is a variant of double outlet right ventricle with concordant atrioventricular connections, a doubly committed ventricular septal defect with absence or deficiency of the conal septum, and unobstructed pulmonary outflow tract.		Double outlet right ventricle with doubly committed interventricular communication without pulmonary stenosis (ventricular septal defect type)	
53	53	01.01.24	Double outlet right ventricle with intact ventricular septum	A congenital cardiovascular malformation that is a variant of double outlet right ventricle that is associated with an intact ventricular septum.			

Table 2. (*Continued*)

54	54	01.05.03	Double outlet left ventricle	A congenital cardiovascular malformation in which both great arteries arise entirely or predominantly from the morphologically left ventricle.			DOLV
55	55	09.01.01	Common arterial trunk	A congenital cardiovascular malformation in which a single arterial trunk arises from the heart, giving origin sequentially to the coronary arteries, one or more pulmonary arteries, and the systemic arterial circulation.	This category includes Collett and Edwards truncus arteriosus types I, II, III and Van Praagh truncus arteriosus types 1, 2, 3, and 4. This category does not include Collett and Edwards truncus arteriosus type IV (which consists of pulmonary atresia with ventricular septal defect and absent intrapericardial pulmonary arteries) or "Tetralogy of Fallot with pulmonary atresia and systemic-to-pulmonary collateral arteries".	Truncus arteriosus communis; Persistent truncus arteriosus; Common truncus arteriosus; Truncus arteriosus	CAT, PTA, TAC, TA
56	56	09.01.15	Common arterial trunk with aortic dominance	A congenital cardiovascular malformation in which a common arterial trunk is associated with an unobstructed aortic arch.			
57	57	09.01.14	Common arterial trunk with aortic dominance and both pulmonary arteries arising from trunk	A congenital cardiovascular malformation in which a common arterial trunk is associated with an unobstructed aortic arch and both pulmonary arteries arise from the common arterial trunk.	This category includes Collett and Edwards truncus arteriosus types I, II, III and Van Praagh truncus arteriosus types 1, 2.		
58	58	09.01.11	Common arterial trunk with aortic dominance and one pulmonary artery absent from the trunk, isolated pulmonary artery	A congenital cardiovascular malformation in which a common arterial trunk is associated with an unobstructed aortic arch and one pulmonary artery does not arise from the common arterial trunk.		Truncus arteriosus with discontinuous pulmonary arteries; Common arterial trunk with discontinuous pulmonary arteries; Van Praagh truncus arteriosus type 3	
59	59	09.01.12	Common arterial trunk with pulmonary dominance and aortic arch obstruction	A congenital cardiovascular malformation in which a common arterial trunk is associated with aortic arch obstruction or interruption.		Common arterial trunk with aortic arch obstruction; Truncus arteriosus with aortic arch obstruction; Van Praagh truncus arteriosus type 4	
60	60	09.01.18	Common arterial trunk with pulmonary dominance and interrupted aortic arch	A congenital cardiovascular malformation in which a common arterial trunk is associated with an interrupted aortic arch.		Truncus arteriosus with interrupted aortic arch; Van Praagh truncus arteriosus type 4	
61	61	09.01.19	Common arterial trunk with pulmonary dominance and aortic coarctation	A congenital cardiovascular malformation in which a common arterial trunk is associated with aortic coarctation.		Truncus arteriosus with coarctation of aorta; Van Praagh truncus arteriosus type 4	

(*Continued*)

Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
62	A^{+}	09.02.10	Atypical truncal valve	A congenital cardiovascular malformation in which the truncal valve does not have the usual morphological or functional attributes at birth. Additional information: this may include truncal valves with more than trivial or mild dysplasia, stenosis, or regurgitation at birth, absent or perforated truncal valve leaflet(s), or a truncal valve that is made up of less than three or more than four leaflets.		Congenital abnormality of truncal valve	
63	62	09.02.19	Congenital truncal valvar regurgitation	A congenital cardiovascular malformation in which there is backward flow through the truncal valve.		Congenital truncal valvar incompetence; Congenital truncal valvar insufficiency; Congenital truncal valve regurgitation; Congenital truncal valve insufficiency; Truncal insufficiency; Truncal regurgitation; Truncal incompetence	
64	63	09.02.18	Congenital truncal valvar stenosis	A congenital cardiovascular malformation in which there is obstruction to flow through the truncal valve due to narrowing or stricture.		Congenital truncal valve stenosis	
65	64	09.02.01	Dysplasia of truncal valve	A congenital cardiovascular malformation where the truncal valve leaflets are markedly thickened with restricted mobility, characterized by the presence of myxomatous tissue.		Congenital truncal valve dysplasia	
66	65	04.00.07	Congenital anomaly of mediastinal vein	A congenital cardiovascular malformation in which there is an abnormality of a mediastinal vein including but not limited to: pulmonary veins, caval veins, coronary sinus, coronary veins, hepatic veins connecting to the heart, brachiocephalic veins, azygos veins, and/or levo-atrial cardinal veins.			

Table 2. (*Continued*)

67	66	04.00.08	Congenital anomaly of mediastinal systemic vein	A congenital cardiovascular malformation in which there is an abnormality of a mediastinal systemic vein including but not limited to: caval veins, coronary sinus, coronary veins, hepatic veins connecting to the heart, brachiocephalic veins, and/or azygos veins.			
68	67	04.01.09	Congenital anomaly of superior caval vein	A congenital cardiovascular malformation in which there is an abnormality of the superior caval vein (superior vena cava).		Congenital anomaly of superior vena cava	Congenital anomaly of SCV; Congenital anomaly of SVC
69	68	04.01.05	Absent right superior caval vein	A congenital cardiovascular malformation in which there is no right superior caval vein (superior vena cava) in the setting of the usual atrial arrangement (atrial situs solitus).		Absent right superior vena cava	Absent RSCV; Absent RSVC
70	69	04.01.25	Left superior caval vein	A congenital cardiovascular malformation in which there is a left superior caval vein (superior vena cava).	Unless the code for absent right superior caval vein is used, this term assumes that a right superior caval vein is present and, therefore, there are bilateral superior caval veins with or without a bridging vein.	Bilateral superior venae cavae; Bilateral superior caval veins; Persistent left superior caval vein; Persistent left superior vena cava	Persistent LSCV, Persistent LSVC, Bilateral SCVs, Bilateral SVCs
71	70	04.01.01	Left superior caval vein to coronary sinus	A congenital cardiovascular malformation in which there is a left superior caval vein (superior vena cava) entering a left-sided coronary sinus.	Unless the code for absent right superior caval vein is used, this term assumes that a right superior caval vein is present and, therefore, there are bilateral superior caval veins with or without a bridging vein.	Persistent left superior caval vein to coronary sinus; Persistent left superior vena cava to coronary sinus	LSCV to CS, LSVC to CS, PLSCV to CS; PLSVC to CS
72	71	04.01.02	Left superior caval vein to left-sided atrium	A congenital cardiovascular malformation in which there is a left superior caval vein (superior vena cava) connecting directly to the left-sided atrium.	Unless the code for absent right superior caval vein is used, this term assumes that a right superior caval vein is present and, therefore, there are bilateral superior caval veins with or without a bridging vein. This term should not be used in the presence of situs inversus of the atria where this is the expected arrangement.	Left superior caval vein persisting to left-sided atrium; Left superior vena cava to left-sided atrium	Persistent LSCV to LA; Persistent LSVC to LA
73	B^{+}	04.01.07	Congenital stenosis of superior caval vein	A congenital cardiovascular malformation in which there is narrowing or stricture of a superior caval vein (superior vena cava).			

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Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
74	72	04.03.08	Congenital anomaly of inferior caval vein	A congenital cardiovascular malformation in which there is an abnormality of the inferior caval vein (inferior vena cava).		Congenital malformation of inferior vena cava; Congenital anomaly of the inferior vena cava	
75	73	04.03.10	Interrupted inferior caval vein with absent suprarenal segment and azygos continuation	A congenital cardiovascular malformation in which there is an absence of the renal-to-hepatic segment of the inferior caval vein (inferior vena cava) with connection to a superior caval vein (superior vena cava) through the azygos venous system.		Interrupted inferior vena cava with absent suprarenal segment and azygos continuation	
76	C^{+}	04.03.06	Congenital stenosis of inferior caval vein	A congenital cardiovascular malformation in which there is narrowing or stricture of the inferior caval vein (inferior vena cava).			
77	74	04.04.05	Congenital anomaly of the coronary sinus	A congenital cardiovascular malformation in which there is an abnormality of the coronary sinus.			
78	75	04.04.13	Unroofed coronary sinus	A congenital cardiac malformation in which there is direct communication between the left atrium and the coronary sinus.	This term includes partial, and complete unroofing or fenestration of the coronary sinus in the presence or absence of an interatrial communication. If an interatrial communication is present through the coronary sinus orifice, then also select the term “interatrial communication through coronary sinus orifice”. If a left superior caval vein (superior vena cava) is present then one should also select the term for “left superior caval vein (superior vena cava) to left-sided atrium”.		
79	D^{+}	04.04.02	Completely unroofed coronary sinus	A congenital cardiac malformation in which there is complete absence of the walls of the coronary sinus and left atrium that usually separate the lumen of the coronary sinus from the left atrial cavity.			
80	E^{+}	04.04.01	Partially unroofed coronary sinus	A congenital cardiac malformation in which there is a communication between the lumen of the coronary sinus and the left atrial cavity.			

Table 2. (*Continued*)

81	76	04.04.14	Coronary sinus orifice atresia or stenosis	A congenital cardiac malformation in which the orifice of the coronary sinus is narrowed or absent.		Coronary sinus ostial atresia or stenosis	
82	77***	04.02.13	Anomalous hepatic venous connection to heart	A congenital cardiovascular malformation in which a hepatic vein or hepatic veins do not connect to the inferior caval vein (inferior vena cava) but connect directly to the heart.	This term does not apply to interruption of the inferior caval vein (inferior vena cava) with a single connection of the hepatic veins to the heart. Coding note: This term in ICD-11 resides in the section entitled "Developmental anomalies of the liver" and is a child of "Congenital anomaly of the hepatic veins".		
83	78	04.08.04	Congenital anomaly of pulmonary vein	A congenital cardiovascular malformation in which there is an abnormality of the pulmonary veins.		Congenital malformation of pulmonary vein	
84	79	04.08.07	Anomalous pulmonary venous connection	A congenital cardiovascular malformation in which one or more pulmonary vein(s) do(es) not connect normally to the morphologically left atrium.			APVC
85	80	04.08.05	Total anomalous pulmonary venous connection	A congenital cardiovascular malformation in which none of the pulmonary veins connect to the morphologically left atrium.		Totally anomalous pulmonary venous connection; Total anomalous pulmonary venous return	TAPVC, TAPVR, TAPVD
86	81	04.06.00	Total anomalous pulmonary venous connection of the supracardiac type	A congenital cardiovascular malformation with total anomalous pulmonary venous connection to the superior caval vein (superior vena cava) or one of its venous tributaries.		Total anomalous pulmonary venous connection Type 1	TAPVC Type I
87	82	04.08.10	Total anomalous pulmonary venous connection of the cardiac type	A congenital cardiovascular malformation with total anomalous pulmonary venous connection to the right atrium directly or to the coronary sinus or to both.		Total anomalous pulmonary venous connection Type 2; Total anomalous pulmonary venous connection, intracardiac	TAPVC Type 2
88	83	04.08.20	Total anomalous pulmonary venous connection of the infracardiac type	A congenital cardiovascular malformation with infradiaphragmatic total anomalous pulmonary venous connection.		Total anomalous pulmonary venous connection Type 3	TAPVC Type 3
89	84	04.08.30	Total anomalous pulmonary venous connection of the mixed type	A congenital cardiovascular malformation with total anomalous pulmonary venous connection at two or more levels (supracardiac, cardiac, or infracardiac).		Total anomalous pulmonary venous connection Type 4	TAPVC Type 4

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Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
90	85	04.07.01	Partial anomalous pulmonary venous connection	A congenital cardiovascular malformation in which one or more (but not all) of the pulmonary veins connect anomalously to the right atrium or to one or more of its venous tributaries and the remaining pulmonary veins connect to the left atrium.		Partially anomalous pulmonary venous connection; Partial anomalous pulmonary venous return	PAPVC. PAPVD, PAPVR
91	86	01.01.16	Partial anomalous pulmonary venous connection of Scimitar type	A congenital cardiovascular malformation with partial anomalous pulmonary venous connection in which some of the pulmonary veins (usually the right pulmonary veins) connect anomalously to the inferior caval vein (inferior vena cava) or to the right atrium at the insertion of the inferior vena cava.		Partial anomalous pulmonary venous return of Scimitar type	
92	87	03.02.23	Scimitar syndrome	A congenital cardiopulmonary malformation with "partial anomalous pulmonary venous connection of Scimitar type" and one or more of the following: hypoplasia of the right lung with bronchial anomalies, dextrocardia, hypoplasia of the right pulmonary artery, lobar lung sequestration, and anomalous systemic arterial supply to the lower lobe of the right lung directly from the aorta or its main branches.		Pulmonary venolobar syndrome	
93	88	04.08.06	Obstructed anomalous pulmonary venous pathway or connection	A congenital cardiovascular malformation in which the pathway of one or more anomalous pulmonary veins is blocked or impeded.		Obstructed anomalous pulmonary venous return	
94	89	04.08.31	Congenital pulmonary venous stenosis or hypoplasia	A congenital cardiovascular malformation with a pathologic narrowing of one or more pulmonary veins including diffuse hypoplasia, long segment focal/ tubular stenosis and/or discrete stenosis.		Congenital pulmonary vein stenosis and/or hypoplasia	

Table 2. (*Continued*)

95	90	04.08.02	Congenital atresia of pulmonary vein	A congenital cardiovascular malformation with atresia of one or more pulmonary veins.		Congenital pulmonary vein atresia	
96	91	05.00.02	Congenital anomaly of an atrium or atrial septum	A congenital cardiovascular malformation in which there is an abnormality of an atrium and/or atrial septum.		Congenital atrial malformation	
97	92	05.07.01	Congenital anomaly of atrial septum	A congenital cardiac malformation in which there is an abnormality of the atrial septum.		Congenital malformation of atrial septum	
98	93	05.06.04	Restrictive interatrial communication or intact atrial septum when an interatrial shunt is physiologically necessary	A cardiac finding in which there is either absence of an interatrial communication or the communication is smaller than is required to permit physiologically adequate interatrial flow.	This is always an abnormal finding prenatally but is a postnatal finding that should only be coded when present prenatally or when associated with other cardiac abnormalities that result in physiological disturbances secondary to the absence of a larger interatrial communication (such as mitral or tricuspid valve atresia).	Restrictive interatrial communication in the presence of an obligatory interatrial shunt; Restrictive interatrial communication or intact atrial septum in the presence of an obligatory interatrial shunt	
99	94	05.03.03	Aneurysm of the atrial septum	A congenital cardiac finding in which the septum primum is abnormally large (redundant) and results in aneurysmal protrusion into one or both atria.		Atrial septal aneurysm	ASA
100	95	05.04.01	Interatrial communication	A congenital cardiac malformation in which there is a hole or pathway between the atrial chambers.	*Although loosely often termed an "atrial septal defect", not all interatrial communications have a defect in the true atrial septum, and so "atrial septal defect" in this context is not a true synonym	Atrial septal defect*; Congenital atrial septal defect; Auricular septal defect; Interatrial septal defect	ASD
101	96**	05.03.01	Patent oval foramen	A congenital cardiovascular finding in which there is a small interatrial communication (or potential communication) confined to the region of the oval fossa (fossa ovalis) characterized by no deficiency of the primary atrial septum (septum primum) and a normal limbus with no deficiency of the septum secundum (superior interatrial fold).		Patent foramen ovale; Open foramen ovale; Open oval foramen; Persistent foramen ovale	PFO
102	97	05.04.02	Atrial septal defect within oval fossa	A congenital cardiac malformation in which there is an interatrial communication confined to the region of the oval fossa (fossa ovalis), most commonly due to a deficiency of the primary atrial septum (septum primum) but deficiency of the septum secundum (superior interatrial fold) may also contribute.		Secundum atrial septal defect; Ostium secundum atrial septal defect	ASD II; OSASD

(*Continued*)

Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
103	98	05.05.00	Sinus venosus defect	A congenital cardiovascular malformation in which there is a caval vein (vena cava) and/or pulmonary vein (or veins) that overrides the atrial septum or the septum secundum (superior interatrial fold) producing an interatrial or anomalous veno-atrial communication.	Although the term sinus venosus atrial septal defect is commonly used, the lesion is more properly termed a sinus venosus communication because, while it functions as an interatrial communication, this lesion is not a defect of the atrial septum.	Sinus venosus atrial septal defect; Sinus venosus communication	
104	99	05.06.02	Common atrium with separate atrioventricular junctions	A congenital cardiovascular malformation in which there is complete or near-complete absence of the interatrial septum.		Single atrium; Atrium communis	
105	100	05.05.03	Interatrial communication through coronary sinus orifice	A congenital cardiovascular malformation in which there is a communication between the left atrium and the coronary sinus allowing interatrial communication through the coronary sinus ostium.	"Interatrial communication through coronary sinus orifice" may or may not be associated with a persistent left superior caval vein (superior vena cava). This occurs in the absence of the coronary sinus (total unroofing of the coronary sinus) or partial unroofing of the coronary sinus.	Coronary sinus atrial septal defect	
106	101	05.01.13	Congenital anomaly of right atrium	A congenital cardiac malformation in which there is an abnormality of the right atrium.			
107	102	05.01.21	Divided right atrium	A congenital cardiac malformation in which the right atrium is partially divided by a large or obstructive Eustachian valve.		Cor triatriatum dexter; Cor triatriatum dextrum; Right cor triatriatum	
108	F^{+}	05.01.04	Chiari network	A congenital cardiac finding in which there is a filamentous, web-like structure originating from the Eustachian valve near the orifice of the inferior caval vein (inferior vena cava) and connecting to variable parts of the right atrium.		Filigreed network of venous valves	
109	103	05.01.06	Left-sided juxtaposition of the atrial appendages	A congenital cardiac malformation in which the right atrial appendage extends from the right atrium, behind the great arteries, to lie adjacent to the left atrial appendage.	This is frequently associated with horizontal orientation of the atrial septum, hypoplasia or atresia of the tricuspid valve, hypoplasia of the right ventricle, transposition of the great arteries or double outlet right ventricle, and subpulmonary or subaortic stenosis.		

Table 2. (*Continued*)

110	104	05.01.12	Congenital giant right atrium	A congenital cardiac malformation in which the right atrium is severely dilated. This is an isolated finding not secondary to abnormalities of the tricuspid valve or right ventricle.		Congenital right atrial aneurysm
111	105	05.02.11	Congenital anomaly of left atrium	A congenital cardiac malformation in which there is an abnormality of the left atrium.		
112	106	05.02.01	Divided left atrium	A congenital cardiac malformation in which there is a partition that divides the left atrium into a posterior chamber that receives some or all of the pulmonary veins and an anterior chamber that communicates with the left atrial appendage and atrioventricular junction (usually the mitral valve).	In differentiating cor triatriatum from supravalvar mitral ring, in cor triatriatum the posterior compartment contains the pulmonary veins while the anterior contains the left atrial appendage and atrioventricular junction (usually the mitral valve); in supravalvar mitral ring, the anterior compartment contains only the mitral valve orifice.	Cor triatriatum sinister; Cor triatriatum sinistrum; Left cor triatriatum
113	107	05.02.04	Right-sided juxtaposition of the atrial appendages	A congenital cardiac malformation in which the left atrial appendage extends from the left atrium, behind the great arteries, to lie adjacent to the right atrial appendage.	This is commonly associated with hypoplasia or atresia of the mitral valve, hypoplasia of the left ventricle, normal (i.e., subpulmonary only) conus and may be seen with atrioventricular septal defect (common atrioventricular canal) or subaortic or aortic stenosis or atresia.	
114	108	06.00.15	Congenital anomaly of an atrioventricular valve or atrioventricular septum	A congenital cardiac malformation in which there is an abnormality of an atrioventricular valve or atrioventricular septum.		
115	109	06.01.11	Congenital anomaly of tricuspid valve	A congenital cardiac malformation in which there is an abnormality of the tricuspid valve.	Tricuspid atresia is listed in the functionally univentricular heart section.	Congenital tricuspid valve anomaly
116	110	06.01.25	Congenital tricuspid regurgitation	A congenital cardiac finding in which there is backward flow through the tricuspid valve.		Congenital tricuspid insufficiency; Congenital tricuspid incompetence; congenital tricuspid valve regurgitation; Congenital tricuspid valve insufficiency; Congenital tricuspid valve incompetence
117	111	06.01.07	Congenital tricuspid valvar stenosis	A congenital cardiovascular malformation of the tricuspid valve in which there is narrowing or stricture (obstruction to flow)		Congenital tricuspid valve stenosis; Congenital stenosis of tricuspid valve; Congenital tricuspid stenosis

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Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
118	112	06.01.04	Tricuspid annular hypoplasia	A congenital cardiac malformation of the tricuspid valve in which there is annular hypoplasia (incomplete development or underdevelopment so that it is abnormally small [below the lower limit of normal adjusted for body size]). Hypoplasia may or may not be associated with stenosis.		Hypoplasia of tricuspid valvar annulus; Hypoplasia of the tricuspid annulus	
119	113	06.01.03	Dysplasia of tricuspid valve	A congenital cardiac malformation of the tricuspid valve, commonly consisting of leaflet thickening and restricted mobility, with normally hinged leaflets.	This diagnosis is not used for patients with Ebstein malformation of tricuspid valve, which is characterized by abnormally hinged tricuspid valve.	Dysplasia of tricuspid valve; tricuspid valvar dysplasia; dysplastic tricuspid valve	
120	114	06.01.09	Straddling tricuspid valve	A congenital cardiac malformation in which the tricuspid subvalvar apparatus has attachments within both ventricles.	This may or may not be associated with valvar overriding.		
121	115	06.01.05	Overriding tricuspid valve	A congenital cardiac malformation in which the tricuspid valve annulus lies in part above both the right and left ventricles.			
122	116	06.01.34	Ebstein malformation of tricuspid valve	A congenital cardiac malformation of the tricuspid valve and right ventricle that is characterized by incomplete delamination of the septal and inferior (posterior) tricuspid valvar leaflets from the myocardium of the right ventricle, and varying degrees of downward (apical) rotational displacement of the functional annulus.	Associated cardiac anomalies include an interatrial communication, the presence of accessory conduction pathways, and varying degrees of right ventricular outflow tract obstruction, including pulmonary atresia. In the setting of discordant atrioventricular and ventriculo-arterial connections ['Congenitally corrected transposition of the great arteries'], 'Ebstein malformation of tricuspid valve' may be present.	Ebstein syndrome; Ebstein anomaly; Ebstein disease; Ebstein anomaly of tricuspid valve; Ebstein's malformation of tricuspid valve; Ebstein; anomaly or syndrome; Tricuspid valve, Ebstein anomaly; Congenital Ebstein deformity of tricuspid valve	
123	G^{+}	06.01.32	Absent tricuspid valve leaflet	A congenital cardiac malformation in which a tricuspid valve leaflet is missing along with its corresponding subvalvar apparatus, thereby leaving a gap between the two other leaflets, particularly at the level of the atrioventricular junction.			

Table 2. (*Continued*)

124	H^+	06.01.36	True cleft of tricuspid valve leaflet	A congenital cardiac malformation in which a leaflet of the tricuspid valve is divided into two parts.		
125	117	06.02.11	Congenital anomaly of mitral valve	A congenital cardiac malformation in which there is an abnormality of the mitral valve.	Mitral atresia is listed in the functionally univentricular heart section.	Congenital mitral valve anomaly
126	118	06.02.25	Congenital mitral regurgitation	A congenital cardiac finding in which there is backward flow through the mitral valve.		Congenital mitral insufficiency; Congenital mitral incompetence; congenital mitral valve regurgitation; Congenital mitral valve insufficiency; Congenital mitral valve incompetence
127	119	06.02.07	Congenital mitral valvar stenosis	A congenital cardiac malformation of the mitral valve in which there is narrowing or stricture of the valvar orifice (obstruction to flow).		Duroziez disease; congenital mitral stenosis; congenital stenosis of mitral valve
128	120	06.02.04	Mitral annular hypoplasia	A congenital cardiac malformation of the mitral valve in which there is annular hypoplasia (incomplete development or underdevelopment so that it is abnormally small [below the lower limit of normal adjusted for body size]). Hypoplasia may or may not be associated with stenosis.		Hypoplasia of mitral valvar annulus; Hypoplasia of the mitral annulus
129	121	06.02.09	Straddling mitral valve	A congenital cardiac malformation in which the mitral subvalvar apparatus has attachments within both ventricles.	This may or may not be associated with valvar overriding.	
130	122	06.02.05	Overriding mitral valve	A congenital cardiac malformation in which the mitral valve annulus lies in part above both the right and left ventricles.		
131	123	06.02.03	Dysplasia of mitral valve	A congenital cardiac malformation that includes any structural abnormality of the mitral valvar leaflet(s), commonly consisting of leaflet thickening and restricted mobility.		Mitral valve dysplasia; Dysplastic mitral valve

(*Continued*)

Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
132	124	05.02.02	Supravalvar or intravalvar mitral ring	A congenital cardiac malformation in which a ridge of tissue is immediately adjacent or integrally attached to the atrial side of the mitral valve.	The intravalvar variant is distinguished from the supravalvar variant because the intravalvar ring is within the funnel of the mitral valvar leaflets. Both the intravalvar variant and the supravalvar variant are differentiated from divided left atrium because in the latter the posterosuperior compartment contains the pulmonary veins while the antero-inferior contains the left atrial appendage (and the atrioventricular valve [usually mitral valve] in the absence of left atrioventricular valvar atresia); in supravalvar or intravalvar mitral ring, the antero-inferior compartment contains only the mitral valve orifice and not the left atrial appendage.	Supravalvar mitral membrane; Membranous supravalvular mitral stenosis; Congenital supravalvar mitral ring; supravalvar mitral stenosis; Supravalvular congenital mitral stenosis	
133	I^{+}	06.02.23	Congenital intravalvar mitral ring	A congenital cardiac malformation in which there is a fibrous shelf-like ridge of tissue adherent to the atrial surface of the mitral valve.			
134	J^{+}	06.02.17	Congenital supravalvar mitral ring	A congenital cardiac malformation in which there is a fibrous shelf-like ridge of tissue located between the mitral valve and the opening to the left atrial appendage.			
135	125	06.02.72	Congenital mitral valvar prolapse	A congenital cardiac malformation of the mitral valve in which one or both leaflets move to the atrial side of the plane of the annulus in systole.		Congenital mitral valve prolapse	Congenital MVP
136	126	06.02.36	True cleft of anterior mitral leaflet	A congenital cardiac malformation of the mitral valve in which the anterior leaflet is divided into two parts.	This term should not be used for the so-called “mitral cleft” of a common atrioventricular valve, as this malformation in a patient with a common atrioventricular valve is actually the zone of apposition between the superior and inferior bridging leaflets, and not a divided valve leaflet.	Isolated cleft of the anterior mitral leaflet (without common atrioventricular junction); True cleft of anterior mitral leaflet (without common atrioventricular junction).	Cleft MV, MV cleft

Table 2. (*Continued*)

137	127	06.02.21	Congenital anomaly of mitral subvalvar apparatus	A congenital cardiac malformation in which the mitral chords, chordal attachments, or papillary muscles are abnormal.		
138	128	06.02.22	Congenital mitral subvalvar stenosis	A congenital cardiac malformation in which there is stenosis (narrowing or stricture of a duct or a canal) of the subvalvar components (chordae tendineae and/or papillary muscles) of the mitral valve. This diagnosis includes mitral stenosis associated with parachute mitral valve, mitral arcade, and hammock mitral valve.		Subvalvar mitral stenosis
139	129	06.02.56	Parachute malformation of mitral valve	A congenital cardiac malformation in which the chords of the mitral valve attach to a single or to closely adjacent papillary muscles.		Parachute mitral valve
140	K^+	06.02.39	Accessory tissue on mitral valve leaflet	A congenital cardiac malformation in which there is extraneous leaflet material forming a string, sheet, or pedunculated mass attached to the atrial or ventricular surface of the mitral valve.		
141	L^+	06.02.32	Congenital unguarded mitral orifice	A congenital cardiac malformation of the mitral valve in which there is a patent valve orifice with complete absence of both leaflets.		
142	M^+	06.02.33	Double orifice of mitral valve	A congenital cardiac malformation of the mitral valve in which there are two separate flow orifices through the mitral valve into the left ventricle.	To differentiate the congenital term “Double orifice of mitral valve” from a mitral valve with a fenestrated or perforated mitral valve leaflet, this term should only be used when both orifices are supported by tensor apparatuses.	
143	130	06.04.11	Congenital anomaly of left-sided atrioventricular valve in double inlet ventricle	A congenital cardiac malformation in association with double inlet ventricle (excluding common atrioventricular valve) in which the atrioventricular valve morphology cannot be determined. This term should be used for the left-sided atrioventricular valve in those hearts.		Left atrioventricular valve stenosis or regurgitation
144	131	06.03.11	Congenital anomaly of right-sided atrioventricular valve in double inlet ventricle	A congenital cardiac malformation in association with double inlet ventricle (excluding common atrioventricular valve) in which the atrioventricular valve morphology cannot be determined. This term should be used for the right-sided atrioventricular valve in those hearts.		

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Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
145	132	06.06.11	Common atrioventricular junction	A congenital cardiac malformation where both atria connect to a common atrioventricular valve which characteristically has 4 or 5 leaflets including superior and inferior bridging leaflets with a single annulus. The common valve may have one or two major orifices depending on the absence or presence of fusion of the bridging leaflets to each other or the septal crest.	The left ventricular zone of apposition between the superior and inferior bridging leaflets is commonly referred to as a "cleft".	Common atrioventricular canal	
146	133	06.06.00	Common atrioventricular junction with atrioventricular septal defect	A congenital cardiac malformation with a common atrioventricular junction and an atrioventricular septal defect.		Atrioventricular septal defect; Atrioventricular canal; Atrioventricular canal defect	AVSD, AVC, AVC defect
147	134	06.07.27	Atrioventricular septal defect with balanced ventricles	A congenital cardiac malformation that is a variant of atrioventricular septal defect (atrioventricular canal defect) with ventricles that are equal or nearly equal in size.		Balanced atrioventricular canal	
148	135	06.07.26	Atrioventricular septal defect with ventricular imbalance	A congenital cardiac malformation that is a variant of atrioventricular septal defect (atrioventricular canal defect) with one ventricle significantly larger than the other.	Unbalanced ventricular size and unbalanced relation of the common atrioventricular valve to the ventricles are to be distinguished by coding unbalanced ventricular size as "Atrioventricular septal defect with ventricular imbalance" and the unbalanced relation of the common atrioventricular valve to the ventricles should also be coded as "Common atrioventricular valve with unbalanced commitment of valve to ventricles".	Unbalanced atrioventricular canal	
149	136	06.07.05	Atrioventricular septal defect with ventricular imbalance with dominant right ventricle and hypoplastic left ventricle	A congenital cardiac malformation that is a variant of an atrioventricular septal defect (atrioventricular canal defect) with the right ventricle significantly larger than the left.		Right dominant AV canal/AVSD	

Table 2. (*Continued*)

150	137	06.07.06	Atrioventricular septal defect with ventricular imbalance with dominant left ventricle and hypoplastic right ventricle	A congenital cardiac malformation that is a variant of an atrioventricular septal defect (atrioventricular canal defect) with the left ventricle significantly larger than the right.	Left dominant AV canal/AVSD	
151	138	06.06.01	Atrioventricular septal defect with communication at the atrial level only	A congenital cardiac malformation that is a variant of an atrioventricular septal defect (atrioventricular canal defect) with an interatrial communication just above the atrioventricular valve, no interventricular communication just below the atrioventricular valve, separate right and left atrioventricular valvar orifices, and varying degrees of malformation of the left-sided component of the common atrioventricular valve. The bridging leaflets of the common atrioventricular valve are bound down to the crest of the scooped-out ventricular septum so that the potential for shunting through the atrioventricular septal defect is possible only at the atrial level and not at the ventricular level.	Incomplete atrioventricular septal defect with isolated atrial component; Incomplete atrioventricular canal defect with isolated atrial component; Ostium primum atrial septal defect; Atrioventricular defect with atrial shunting only; Primum atrial septal defect; Partial atrioventricular canal defect with isolated atrial component; Partial atrioventricular septal defect: ostium primum type; Partial atrioventricular septal defect	PAVSD, PAVC
152	139	06.06.08	Atrioventricular septal defect with communication at the ventricular level only	A congenital cardiac malformation that is a variant of an atrioventricular septal defect (atrioventricular canal defect) with an interventricular communication just below the atrioventricular valve, no interatrial communication just above the atrioventricular valve, separate right and left atrioventricular valvar orifices, and varying degrees of malformation of the left-sided component of the common atrioventricular valve. The bridging leaflets of the common atrioventricular valve are bound to the atrial septum so that the potential for shunting through the atrioventricular septal defect is possible only at the ventricular level and not at the atrial level.	AV canal/AVSD with isolated VSD; Atrioventricular canal defect with isolated ventricular communication	

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Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
153	140	06.06.10	Atrioventricular septal defect with communication at atrial level and restrictive communication at ventricular level	A congenital cardiac malformation that is a variant of an atrioventricular septal defect (atrioventricular canal defect) with an interatrial communication immediately above the atrioventricular valve, and a restrictive interventricular communication immediately below the atrioventricular valve.	This term is used to identify hearts with a restrictive interventricular communication. If there is a single atrioventricular valvar orifice this term should still be used. If the ventricular component of the atrioventricular septal defect is unrestrictive (no interventricular pressure gradient), this malformation should not be coded as "Atrioventricular septal defect (atrioventricular canal defect) with communication at atrial level and restrictive communication at ventricular level" and instead the term "Atrioventricular septal defect (atrioventricular canal defect) with communication at atrial level and unrestrictive communication at ventricular level" should be used.	Intermediate atrioventricular septal defect with atrial and ventricular components and separate atrioventricular valvar orifices; Atrioventricular canal defect associated with a restrictive ventricular septal defect; Atrioventricular septal defect with atrial shunting and restrictive ventricular shunting; Intermediate atrioventricular canal defect with atrial and ventricular components and separate atrioventricular valve; Transitional atrioventricular canal defect with atrial and ventricular components and separate atrioventricular valve; Transitional atrioventricular septal defect with atrial and ventricular components and separate atrioventricular valves	
154	141	06.06.09	Atrioventricular septal defect with communication at atrial level and unrestrictive communication at ventricular level	A congenital cardiac malformation that is a variant of an atrioventricular septal defect (atrioventricular canal defect) with an interatrial communication just above the atrioventricular valve, an interventricular communication just below the atrioventricular valve, and varying degrees of malformation of the left ventricular component of the common atrioventricular valve. There is unrestrictive interventricular communication (no interventricular pressure gradient) and the bridging leaflets usually float to varying extent within the atrioventricular septal defect.		Complete atrioventricular septal defect; Complete atrioventricular canal defect	CAVSD, CAVC
155	142	01.01.20	Atrioventricular septal defect and tetralogy of Fallot	A congenital cardiac malformation with both an atrioventricular septal defect (atrioventricular canal defect) and tetralogy of Fallot.	Tetralogy of Fallot with atrioventricular septal defect (common atrioventricular canal) is always the complete form (unrestrictive interventricular component) and usually has four leaflets of the common atrioventricular valve (undivided superior leaflet) and very few or no attachments of that leaflet to the crest of the ventricular septum.	Atrioventricular septal defect associated with tetralogy of Fallot; Atrioventricular canal and Tetralogy of Fallot	TOF/AVC, TOF/AVSD

Table 2. (*Continued*)

156	143	05.06.03	Common atrium with common atrioventricular junction	A congenital cardiac malformation in which there is complete absence of the interatrial septum in the setting of a common atrioventricular junction (common atrioventricular canal).		Single atrium
157	144	06.05.60	Common atrioventricular valvar regurgitation	A congenital cardiac finding where there is backward flow through the common atrioventricular valve.		
158	145	06.05.14	Atypical common atrioventricular valve	A congenital cardiac malformation in which the common atrioventricular valve does not have the usual morphology of 4-5 non-dysplastic leaflets that relate relatively equally to both ventricles.		
159	146	06.07.36	Common atrioventricular valve with unbalanced commitment of valve to ventricles	A congenital cardiac malformation in which the common atrioventricular valve is primarily related to one ventricle, usually but not always associated with hypoplasia of the other ventricle.	Unbalanced ventricular size and unbalanced relation of the common atrioventricular valve to the ventricles are to be distinguished by coding unbalanced ventricular size as “Atrioventricular septal defect with ventricular imbalance” and the unbalanced relation of the common atrioventricular valve to the ventricles should also be coded as “Common atrioventricular valve with unbalanced commitment of valve to ventricles”.	
160	147	06.07.37	Common atrioventricular valve with unbalanced commitment of valve to right ventricle	A congenital cardiac malformation in which the common atrioventricular valve is primarily related to the right ventricle, usually but not always associated with hypoplasia of the left ventricle.		
161	148	06.07.38	Common atrioventricular valve with unbalanced commitment of valve to left ventricle	A congenital cardiac malformation in which the common atrioventricular valve is primarily related to the left ventricle, usually but not always associated with hypoplasia of the right ventricle.		
162	149	06.05.71	Atypical right ventricular component of common atrioventricular valve	A congenital cardiac malformation in which the right ventricular component of the common atrioventricular valve does not have the usual structure (independently supported and well developed right ventricular components of the common atrioventricular valve).		

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Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
163	150	06.05.72	Atypical left ventricular component of common atrioventricular valve	A congenital cardiac malformation in which the left ventricular component of the common atrioventricular valve does not have the usual structure (independently supported and well developed left ventricular components of the common atrioventricular valve).			
164	151	06.05.25	Double orifice of left ventricular component of common atrioventricular valve	A congenital cardiac malformation in which the left ventricular component of the common atrioventricular valve has two orifices.			
165	152	06.05.98	Deficient mural leaflet of left ventricular component of common atrioventricular valve	A congenital cardiac malformation in which the left ventricular component of the common atrioventricular valve is composed primarily of the bridging leaflets due to hypoplasia or absence of the mural (lateral) leaflet.		Deficient lateral leaflet of left ventricular component of common atrioventricular valve	
166	153	06.07.28	Common atrioventricular junction without an atrioventricular septal defect	A congenital cardiac malformation in which there is a common atrioventricular junction without any communication at the level of the atrioventricular septum.	This code could be used when there is a trifoliate left atrioventricular valve, commonly known as cleft mitral valve secondary to atrioventricular septal defect, and also has evidence of spontaneous obliteration of the atrioventricular septal defect by valvar or subvalvar tissue. Additional defects in the atrial or ventricular septums that do not involve the atrioventricular septum are not excluded by this term and should be coded separately.	Common atrioventricular junction with spontaneous fibrous closure of atrioventricular septal defect; Common atrioventricular junction with intact atrioventricular septal structures	
167	154	07.14.02	Communication between left ventricle and right atrium	A deficiency of the atrioventricular component of the membranous septum permitting shunting of blood from the left ventricle to the right atrium.	This is an isolated absence of the atrioventricular septum without other components that may be commonly seen in association with spectrum of disorders associated with atrioventricular septal defects such as a common atrioventricular valve or interatrial or interventricular communications. The Gerbode defect may be confused with ventricular septal defects where a deficiency of tricuspid valvar tissue permits ventriculo-atrial shunting subsequent to initial interventricular shunting (left ventricle to right ventricle to right atrium).	Gerbode defect	

Table 2. (*Continued*)

168	155	07.00.00	Congenital anomaly of a ventricle or the ventricular septum	A congenital cardiac malformation in which there is an abnormality of a ventricle and/or the ventricular septum. The ventricles include the ventricular inlet, ventricular body and ventricular outflow tract.		Congenital ventricular or ventricular septal malformation	
169	156	07.01.07	Congenital right ventricular anomaly	A congenital cardiac malformation in which there is an abnormality of the right ventricle.			
170	157	07.02.00	Right ventricular hypoplasia	A congenital cardiac malformation in which the right ventricle is abnormally small (below the lower limit of normal adjusted for body size).	This morphological abnormality usually is an integral part of other congenital cardiovascular anomalies and, therefore, frequently does not need to be coded separately. It should, however, be coded as secondary to an accompanying congenital cardiovascular anomaly if the right ventricular hypoplasia is not considered an integral and understood part of the primary congenital cardiovascular diagnosis. It would rarely be coded as a primary and/or isolated diagnosis.	Hypoplastic right ventricle	
171	158	07.05.20	Congenital right ventricular outflow tract obstruction	A congenital cardiac condition in which the flow through the right ventricular outflow tract (proximal to the valve[s] guarding the outflow from the right ventricle) is blocked or impeded.			RVOTO
172	159	07.03.01	Double chambered right ventricle	A congenital cardiac malformation in which the right ventricle is divided into two chambers, one inferior including the inlet and trabecular portions of the right ventricle and one superior including the trabecular portion and infundibulum.	Double chamber right ventricle is often associated with one or several closing ventricular septal defects. In some cases, the ventricular septal defect is already closed. Double chamber right ventricle is differentiated from the rare isolated infundibular stenosis that develops more superiorly.	Double-chamber right ventricle; Anomalous right ventricular muscle bundle; Double chamber right ventricle	DCRV
173	160	07.01.13	Right ventricular myocardial sinusoids	A congenital cardiac malformation in which there are deep, endothelial lined, blind-ended intramyocardial tunnels communicating with the right ventricular chamber. These occur in the setting of pulmonary atresia or severe right ventricular outflow tract obstruction, intact ventricular septum and a patent tricuspid valve.	These RV sinusoids do not communicate with the coronary arteries. Where connections from the right ventricle to the coronary arteries exist, one should code for "Congenital coronary artery-to-right ventricular fistula".		

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Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
174	161	07.01.06	Parchment right ventricle	A congenital cardiac malformation in which there is almost complete absence of right ventricular myocardium and preserved septal and left ventricular myocardium.	Parchment right ventricle can occur in association with a normal tricuspid valve (Uhl anomaly) or with an abnormal tricuspid valve, such as Ebstein Anomaly. This should be distinguished from arrhythmogenic right ventricular cardiomyopathy by the presence of fatty infiltration in the latter.	Uhl's anomaly; Uhl anomaly	
175	162	01.01.01	Tetralogy of Fallot	A group of congenital cardiac malformations with biventricular atrioventricular alignments or connections characterized by anterosuperior deviation of the conal or outlet septum or its fibrous remnant, narrowing or atresia of the pulmonary outflow, a ventricular septal defect of the malalignment type, and biventricular origin of the aorta. Tetralogy of Fallot will always have a ventricular septal defect, narrowing or atresia of the pulmonary outflow, aortic override, and most often right ventricular hypertrophy.		Tetrad of Fallot; Fallot tetralogy; Fallot tetrad; Fallot disease; Fallot complex; Fallot's tetralogy; Subpulmonic stenosis, ventricular septal defect, overriding aorta, and right ventricular hypertrophy; Ventricular septal defect with pulmonary stenosis or atresia, dextroposition of aorta, and hypertrophy of right ventricle	TOF
176	163	09.05.25	Tetralogy of Fallot with absent pulmonary valve syndrome	A congenital cardiovascular malformation that is a variant of tetralogy of Fallot in which the ventriculo-arterial junction of the right ventricle with the pulmonary trunk features an atypical valve with absent or rudimentary leaflets (cusps) that do not coapt. In its usual form there is dilatation of the pulmonary trunk and central right and left pulmonary arteries, which when extreme, is associated with abnormal arborization of lobar and segmental pulmonary artery branches and with compression of the trachea and mainstem bronchi, often with tracheobronchomalacia.	A congenital cardiovascular malformation that is a variant of tetralogy of Fallot in which the ventriculo-arterial junction of the right ventricle with the pulmonary trunk features an atypical valve with absent or rudimentary leaflets (cusps) that do not coapt. In its usual form there is dilatation of the pulmonary trunk and central right and left pulmonary arteries, which when extreme, is associated with abnormal arborization of lobar and segmental pulmonary artery branches and with compression of the trachea and mainstem bronchi, often with tracheobronchomalacia. The physiologic consequence is usually a combination of variable degrees of both stenosis and regurgitation of the pulmonary valve.		

Table 2. (*Continued*)

177	164	01.01.26	Tetralogy of Fallot with pulmonary atresia	A congenital cardiovascular malformation that is a variant of tetralogy of Fallot in which there is no direct communication between the right ventricle and the pulmonary arterial tree.	This term should not be used when major systemic-to-pulmonary artery collaterals (including major aortopulmonary collateral arteries [MAPCAs]) are known to be present.	Pulmonary atresia with ventricular septal defect [Fallot type]
178	165	01.01.57	Tetralogy of Fallot with pulmonary atresia and systemic-to-pulmonary collateral arteries	A congenital cardiovascular malformation that is a variant of tetralogy of Fallot in which there is no direct communication between the right ventricle and the pulmonary arterial tree and there are collateral blood vessels between the systemic and pulmonary arteries.		Pulmonary atresia with ventricular septal defect and systemic-to-pulmonary collateral artery(ies) [Fallot type]; Pulmonary atresia, ventricular septal defect and MAPCAs
179	166	07.06.07	Congenital left ventricular anomaly	A congenital cardiac malformation in which there is an abnormality of the left ventricle.		
180	167	07.07.00	Left ventricular hypoplasia	A congenital cardiac malformation in which the left ventricle is abnormally small (below the lower limit of normal adjusted for body size).	This morphological abnormality usually is an integral part of other congenital cardiovascular anomalies and does not need to be coded separately if this is the case. It should be coded as secondary to an accompanying congenital cardiovascular anomaly if the left ventricular hypoplasia is not considered an integral and understood part of the primary congenital cardiovascular diagnosis such as hypoplastic left heart syndrome.	Hypoplastic left ventricle
181	168	07.06.19	Congenital left ventricular aneurysm or diverticulum	A congenital cardiac malformation in which there is an outpouching of the left ventricular wall.		
182	N^{+}	07.06.01	Congenital left ventricular aneurysm	A congenital cardiac malformation in which there is a discrete region of the ventricular wall that is thinner than the adjacent myocardial segments, bulges away from the lumen of the left ventricle during systole, and exhibits either akinesis or dyskinesis.		
183	O^{+}	07.06.03	Congenital left ventricular diverticulum	A congenital cardiac malformation in which there is an outpouching from the ventricular wall that contains endocardium, myocardium, and pericardium and displays normal contraction.		

(*Continued*)

Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
184	169	07.09.28	Congenital left ventricular outflow tract obstruction	A congenital cardiac condition in which the flow through the left ventricular outflow tract (proximal to the valve[s] guarding the outflow from the left ventricle) is blocked or impeded.	This code should not be used for obstruction immediately under the arterial valve such as subaortic stenosis due to fibromuscular shelf or tunnel.		LVOTO
185	170	07.09.08	Congenital left ventricular outflow tract obstruction due to atrioventricular valve	A congenital cardiac malformation in which the flow through the left ventricular outflow tract is blocked or impeded due to abnormal position or attachments of tricuspid, mitral, or common atrioventricular valvar tissue.			
186	171	01.01.33	Left heart obstruction at multiple sites	A congenital cardiovascular malformation in which more than one of the following lesions are present: (1) supravalvar or intravalvar mitral ring, (2) mitral subvalvar stenosis, (3) a parachute deformity of the mitral valve, (4) subaortic stenosis, (5) valvar aortic stenosis, and (6) aortic coarctation.	Shone's syndrome consists of multilevel hypoplasia or obstruction of the left-heart. The syndrome is based on the original report from Shone that was based on analysis of 8 autopsied cases and described the tendency of these four obstructive, or potentially obstructive, conditions to coexist. Only 2 of the 8 cases exhibited all four conditions, with the other cases exhibiting only two or three of the anomalies.	Shone syndrome; Shone's syndrome; Shone's anomaly; Shone's complex; Shone's disease	
187	172	07.06.12	Left ventricular myocardial sinusoids	A congenital cardiac malformation in which there are deep, endothelial lined, blind-ended intramyocardial tunnels communicating with the left ventricular chamber. These occur in the setting of aortic atresia or severe left ventricular outflow tract obstruction, intact ventricular septum and a patent mitral valve.	These left ventricular sinusoids do not communicate with the coronary arteries. Where connections from the left ventricle to the coronary arteries exist, one should code for "Congenital coronary artery-to-left ventricular fistula". This code should not be used for left ventricular non-compaction.		
188	P^{+}	07.00.07	Anomalous ventricular bands	A congenital cardiac malformation in which fibromuscular tissue crosses through a ventricular cavity, inserting at two or more points on the ventricular wall or papillary muscles.			
189	173	07.20.04	Congenital anomaly of ventricular septum	A congenital cardiac malformation in which there is an abnormality of the interventricular septum.		• anomaly; ventricular septum • ventricle septa anomaly • ventricle septal heart anomaly • ventricle septum heart anomaly • ventricular septa anomaly • ventricular septal heart anomaly	

Table 2. (*Continued*)

190	174	07.14.07	Restrictive interventricular communication when an interventricular shunt is physiologically necessary	A congenital cardiac finding in which there is an interventricular communication that is smaller than is required to permit physiologically adequate interventricular flow.	This should be coded only when associated with other cardiac abnormalities that result in physiological disturbances secondary to the absence of a larger interventricular communication. Clinically important examples include double outlet right ventricle or tetralogy of Fallot with restrictive ventricular septal defect, double inlet left ventricle or tricuspid atresia with concordant or discordant great arterial connections and restrictive ventricular septal defect, and others.		
191	175	07.10.00	Ventricular septal defect	A congenital cardiac malformation in which there is a hole or pathway between the ventricular chambers.	The definitions offered for a "ventricular septal defect", in its various forms, will be used most frequently in the setting of patients who do not have abnormalities of either the atrioventricular or ventriculo-arterial connections. The definitions themselves, however, are equally applicable for the description and categorization of holes or pathways between the ventricles when the segmental connections between the cardiac components are abnormal. The key to understanding the definitions is to appreciate that the hole or pathway between the ventricles is defined both on the basis of its geographic location within the ventricular septum and its margins as seen from the aspect of the morphologically right ventricle. In this respect, when the ventriculo-arterial connections are concordant or discordant, the roof of the channel, as viewed from the right ventricle, is the muscular outlet septum or its fibrous remnant. When both arterial trunks arise from the morphologically right ventricle, in contrast, the channel between the ventricles provides the outflow from the morphologically left ventricle, and is roofed by the inner heart curvature. In the setting of double outlet right ventricle, therefore, the interventricular	Interventricular communication; Interventricular septal defect; Congenital ventricular septal defect	VSD

(*Continued*)

Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
					communication is not the same geometric locus as the ventricular septal defect, although many still continue to describe the outlet from the left ventricle as the "ventricular septal defect". In univentricular atrioventricular connections with functionally single left ventricle with an outflow chamber, the communication, which used to be called a bulboventricular foramen, is both a ventricular septal defect and an interventricular communication. The situation in the setting of double outlet right ventricle, however, shows that the terms cannot always be used as being synonymous. A similar situation pertains in the setting of common arterial trunk, since the hole usually closed by the surgeon to provide septal integrity is the right ventricular margin of the channel between the ventricles, rather than the geometric interventricular communication.		
192	176	07.10.01	Perimembranous central ventricular septal defect	A congenital cardiovascular malformation in which there is a ventricular septal defect that 1) occupies the space that is usually closed by the interventricular part of the membranous septum, 2) is adjacent to the area of fibrous continuity between the leaflets of an atrioventricular valve and an arterial valve, 3) is adjacent to an area of mitral-tricuspid fibrous continuity, and 4) is located at the center of the base of the ventricular mass.	Although best used to describe the perimembranous defect that opens centrally at the base of the right ventricle, this term might be used to code perimembranous defects with inlet or outlet extension. It is recommended, however, that the more precise terms be used whenever possible for coding the latter lesions. This code is used by some as synonymous with the perimembranous, conoventricular, Type II, or the paramembranous defects. It should not be used to code an inlet VSD, or the so-called atrioventricular canal VSD. More specific terms exist for coding these entities. It is used by some to describe an isolated perimembranous VSD without extension, although it is unlikely	Perimembranous ventricular septal defect; Membranous ventricular septal defect; Paramembranous ventricular septal defect; Type 2 ventricular septal defect; Central perimembranous ventricular septal defect	PMVSD

Table 2. (*Continued*)

					that perimembranous defects exist in the absence of deficiency of their muscular perimeter. The conoventricular VSD with malalignment should be coded as an outlet defect, as should the perimembranous defect opening to the outlet of the right ventricle. All perimembranous defects, nonetheless, have part of their margins made up of fibrous continuity either between the leaflets of an atrioventricular and an arterial valve or, in the setting of double outlet right ventricle or overriding of the tricuspid valve, by fibrous continuity between the leaflets of the mitral and tricuspid valves. Such defects can also extend to become doubly committed and juxta-arterial (conal septal hypoplasia) when there is also fibrous continuity between the leaflets of the arterial valves or when there is a common arterial valve. Specific codes exist for these variants, which ideally should not be coded using this term.	
193	177	07.14.05	Inlet ventricular septal defect without a common atrioventricular junction	A congenital cardiac malformation in which there is a ventricular septal defect that opens predominantly into the inlet component of the right ventricle in the absence of a common atrioventricular junction.		Type 3 ventricular septal defect; Inlet atrioventricular canal type ventricular septal defect; Inlet ventricular septal defect
194	178	07.10.02	Inlet perimembranous ventricular septal defect without atrioventricular septal malalignment without a common atrioventricular junction	A congenital cardiac malformation in which there is a ventricular septal defect that usually, but not always, extends beneath the full annular length of the septal leaflet of the tricuspid valve and to the membranous septum, into the inlet component of the right ventricle in the absence of atrioventricular septal malalignment and of a common atrioventricular junction.		Perimembranous ventricular septal defect with extension to right ventricular inlet (posterior); Inlet perimembranous ventricular septal defect; Perimembranous inlet ventricular septal defect

(*Continued*)

Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
195	179	07.14.06	Inlet perimembranous ventricular septal defect with atrioventricular septal malalignment and without a common atrioventricular junction	A congenital cardiac malformation in which there is an inlet ventricular septal defect with malalignment between the atrial septum and the postero-inferior part of the muscular ventricular septum, in the absence of a common atrioventricular junction, such that there is overriding with or without straddling of the tricuspid valve.		Perimembranous ventricular septal defect with extension to right ventricular inlet (posterior) and atrioventricular septal malalignment; Inlet perimembranous ventricular septal defect with atrioventricular septal malalignment; Perimembranous inlet ventricular septal defect with atrioventricular septal malalignment	
196	180	07.11.02	Inlet muscular ventricular septal defect	A congenital cardiac malformation in which there is a ventricular septal defect with exclusively muscular borders that opens into the inlet component of the right ventricle.		Muscular ventricular septal defect opening into right ventricular inlet; Inlet muscular ventricular septal defect	
197	181	07.11.01	Trabecular muscular ventricular septal defect	A congenital cardiac malformation in which there is a ventricular septal defect within the trabeculated component of the ventricular septum.	The code specifying defects within the trabecular part of the ventricular septum should not be used to code the inlet or outlet muscular defects as there are specific codes for these.	Type 4 ventricular septal defect; Trabecular ventricular septal defect; Muscular trabecular ventricular septal defect	
198	182	07.11.04	Trabecular muscular ventricular septal defect midseptal	A congenital cardiac malformation in which there is a ventricular septal defect that opens to the right ventricle in the middle of the trabeculated component of the ventricular septum.		Midmuscular ventricular septal defect; Trabecular muscular ventricular septal defect: mid; Muscular trabecular ventricular septal defect: Midseptal	
199	183	07.11.03	Trabecular muscular ventricular septal defect apical	A congenital cardiac malformation in which there is a ventricular septal defect that opens to the right ventricle in the apical region of the trabeculated component of the ventricular septum.		Apical muscular ventricular septal defect; Muscular trabecular ventricular septal defect: Apical	
200	184	07.11.12	Trabecular muscular ventricular septal defect postero-inferior	A congenital cardiac malformation in which there is a ventricular septal defect that opens to the right ventricle in the postero-inferior region of the trabeculated component of the ventricular septum.	It can be arbitrary as to when an inlet muscular defect becomes an inferior apical defect. The distinction should be made on whether or not the defect is partly shielded by the septal leaflet of the tricuspid valve.	Postero-inferior muscular ventricular septal defect; Posterior muscular ventricular septal defect; Inferior muscular ventricular septal defect; Muscular ventricular septal defect in postero-inferior trabecular septum; Muscular trabecular ventricular septal defect: postero-inferior	

Table 2. (*Continued*)

201	185	07.11.07	Trabecular muscular ventricular septal defect anterosuperior	A congenital cardiac malformation in which there is a muscular ventricular septal defect that opens to the right ventricle anterior to the body of the septomarginal trabeculation, or septal band.	Anterosuperior muscular ventricular septal defect; Anterior muscular ventricular septal defect; Superior muscular ventricular septal defect; Muscular ventricular septal defect opening anterior to body of septomarginal trabeculation; Muscular trabecular ventricular septal defect: Anterosuperior
202	186	07.11.05	Multiple trabecular muscular ventricular septal defects	A congenital cardiac malformation in which there are multiple muscular ventricular septal defects that open to the right ventricle in the trabeculated component of the ventricular septum.	Multiple muscular trabecular ventricular septal defects; Muscular trabecular ventricular septal defect: Multiple (“Swiss cheese” septum)
203	187	07.12.00	Outlet ventricular septal defect	A congenital cardiac malformation in which there is a ventricular septal defect that opens to the outlet of the right ventricle between or above the limbs of the septal band.	Supracristal ventricular septal defect; Conal septal defect; Infundibular ventricular septal defect; Type 1 ventricular septal defect; Subpulmonary ventricular septal defect; Subarterial ventricular septal defect; Juxtaarterial ventricular septal defect; Conal ventricular septal defect; Conoseptal hypoplasia; Intraconal ventricular septal defect
204	188	07.12.09	Outlet ventricular septal defect without malalignment	A congenital cardiac malformation in which there is an outlet ventricular septal defect, in the absence of malalignment of the outlet septum with the trabecular muscular septum.	
205	189	07.11.06	Outlet muscular ventricular septal defect without malalignment	A congenital cardiac malformation in which there is an outlet ventricular septal defect with exclusively muscular borders, in the absence of malalignment of the outlet septum with the trabecular muscular septum.	Muscular outlet ventricular septal defect with aligned outlet septum; Conal septal hypoplasia with exclusively muscular borders; Muscular outlet ventricular septal defect
206	190	07.12.01	Doubly committed juxta-arterial ventricular septal defect without malalignment	A congenital cardiac malformation in which there is an outlet ventricular septal defect bordered superiorly by the area of fibrous continuity or a fibrous outlet septum between the aortic and pulmonary valves, that is aligned with the trabecular muscular septum.	Doubly committed subarterial ventricular septal defect with aligned outlet septum; Conoseptal hypoplasia with aligned outlet septum; Doubly committed juxta-arterial outlet ventricular septal defect; Doubly committed subarterial outlet ventricular septal defect

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Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
207	191	07.12.02	Doubly committed juxta-arterial ventricular septal defect without malalignment and with muscular postero-inferior rim	A congenital cardiac malformation in which there is an outlet ventricular septal defect with muscular postero-inferior rim, bordered superiorly by the area of fibrous continuity or a fibrous outlet septum between the aortic and pulmonary valves, that is aligned with the trabecular muscular septum.		Doubly committed juxta-arterial ventricular septal defect with muscular postero-inferior rim; Doubly committed subarterial ventricular septal defect with muscular postero-inferior rim	
208	192	07.12.03	Doubly committed juxta-arterial ventricular septal defect without malalignment and with perimembranous extension	A congenital cardiac malformation in which there is an outlet ventricular septal defect with perimembranous extension, bordered superiorly by the area of fibrous continuity or a fibrous outlet septum between the aortic and pulmonary valves, that is aligned with the trabecular muscular septum.		Doubly committed subarterial ventricular septal defect with aligned outlet septum with perimembranous extension; Conal septal hypoplasia without cranial muscular border with aligned outlet septum and with perimembranous extension; Doubly committed juxta-arterial outlet ventricular septal defect with perimembranous extension; Doubly committed juxta-arterial ventricular septal defect with without malaligned fibrous outlet septum and fibrous postero-inferior rim (perimembranous)	
209	193	07.10.17	Outlet ventricular septal defect with anteriorly malaligned outlet septum	A congenital cardiac malformation in which there is an outlet ventricular septal defect and the muscular outlet septum is malaligned in an antero-cranial fashion with respect to the trabecular muscular septum such that there is overriding of the arterial valve supported predominantly by the left ventricle.		Ventricular septal defect opening into right ventricular outlet with anterior malalignment of outlet septum; Outlet ventricular septal defect “Fallot type”	
210	194	07.11.15	Outlet muscular ventricular septal defect with anteriorly malaligned outlet septum	A congenital cardiac malformation in which there is an outlet ventricular septal defect with exclusively muscular borders, and the muscular outlet septum is malaligned in an antero-cranial fashion with respect to the trabecular muscular septum such that there is overriding of the arterial valve supported predominantly by the left ventricle.		Muscular ventricular septal defect opening into right ventricular outlet with anterior malalignment of outlet septum; Muscular outlet ventricular septal defect “Fallot type”; Muscular outlet ventricular septal defect with anteriorly malaligned outlet septum	

Table 2. (*Continued*)

211	195	07.10.04	Outlet perimembranous ventricular septal defect with anteriorly malaligned outlet septum	A congenital cardiac malformation in which there is an outlet ventricular septal defect with perimembranous extension and the muscular outlet septum is malaligned in an antero-cranial fashion with respect to the trabecular muscular septum such that there is overriding of the arterial valve supported predominantly by the left ventricle.	Perimembranous ventricular septal defect with outlet extension and anterior malalignment of outlet septum; Perimembranous outlet ventricular septal defect "Fallot type"; Perimembranous outlet ventricular septal defect with anteriorly malaligned outlet septum
212	196	07.12.12	Doubly committed juxta-arterial ventricular septal defect with anteriorly malaligned fibrous outlet septum	A congenital cardiac malformation in which there is an outlet ventricular septal defect bordered superiorly by the area of fibrous continuity or a fibrous outlet septum between the aortic and pulmonary valves, and the fibrous outlet septum is malaligned in an antero-cranial fashion with respect to the trabecular muscular septum such that there is overriding of the arterial valve supported predominantly by the left ventricle.	Doubly committed subarterial ventricular septal defect and anterior malalignment of the outlet septum; Doubly committed juxta-arterial ventricular septal defect "Fallot type"
213	197	07.12.07	Doubly committed juxta-arterial ventricular septal defect with anteriorly malaligned fibrous outlet septum and muscular postero-inferior rim	A congenital cardiac malformation in which there is an outlet ventricular septal defect with muscular postero-inferior rim, bordered superiorly by the area of fibrous continuity or a fibrous outlet septum between the aortic and pulmonary valves, and the fibrous outlet septum is malaligned in an antero-cranial fashion with respect to the trabecular muscular septum such that there is overriding of the arterial valve supported predominantly by the left ventricle.	Doubly committed subarterial ventricular septal defect with muscular posterior inferior rim and anterior malalignment of outlet septum; Doubly committed juxta-arterial ventricular septal defect with muscular posterior inferior rim "Fallot type"
214	198	07.12.05	Doubly committed juxta-arterial ventricular septal defect with anteriorly malaligned fibrous outlet septum and perimembranous extension	A congenital cardiac malformation in which there is an outlet ventricular septal defect with perimembranous extension, bordered superiorly by the area of fibrous continuity or a fibrous outlet septum between the aortic and pulmonary valves, and the fibrous outlet septum is malaligned in an antero-cranial fashion with respect to the trabecular muscular septum such that there is overriding of the arterial valve supported predominantly by the left ventricle.	Doubly committed subarterial ventricular septal defect with perimembranous extension and anterior malalignment of outlet septum; Doubly committed juxta-arterial ventricular septal defect with perimembranous extension "coarctation type"; Doubly committed juxta-arterial ventricular septal defect with anteriorly malaligned fibrous outlet septum and fibrous postero-inferior rim (perimembranous)

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Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
215	199	07.10.18	Outlet ventricular septal defect with posteriorly malaligned outlet septum	A congenital cardiac malformation in which there is an outlet ventricular septal defect and the muscular outlet septum is malaligned in a postero-caudal fashion with respect to the trabecular muscular septum such that there is usually obstruction to the left ventricular outflow tract.		Ventricular septal defect opening into right ventricular outlet with posterior malalignment of outlet septum; Outlet ventricular septal defect “coarctation type”	
216	200	07.11.16	Outlet muscular ventricular septal defect with posteriorly malaligned outlet septum	A congenital cardiac malformation in which there is an outlet ventricular septal defect with exclusively muscular borders, and the muscular outlet septum is malaligned in a postero-caudal fashion with respect to the trabecular muscular septum such that there is usually obstruction to the left ventricular outflow tract.		Muscular ventricular septal defect opening into right ventricular outlet with posterior malalignment of outlet septum; Muscular outlet ventricular septal defect “coarctation type”; Muscular outlet ventricular septal defect with posteriorly malaligned outlet septum	
217	201	07.10.19	Outlet perimembranous ventricular septal defect with posteriorly malaligned outlet septum				
218	202	07.12.13	Doubly committed juxta-arterial ventricular septal defect with posteriorly malaligned fibrous outlet septum	A congenital cardiac malformation in which there is an outlet ventricular septal defect bordered superiorly by the area of fibrous continuity or a fibrous outlet septum between the aortic and pulmonary valves, and the fibrous outlet septum is malaligned in a postero-caudal fashion with respect to the trabecular muscular septum such that there is usually obstruction to the left ventricular outflow tract.		Doubly committed subarterial ventricular septal defect with posterior malalignment of outlet septum; Doubly committed juxta-arterial ventricular septal defect “coarctation type”	
219	203	07.12.08	Doubly committed juxta-arterial ventricular septal defect with posteriorly malaligned fibrous outlet septum and muscular postero-inferior rim	A congenital cardiac malformation in which there is an outlet ventricular septal defect with muscular postero-inferior rim, bordered superiorly by the area of fibrous continuity or a fibrous outlet septum between the aortic and pulmonary valves, and the fibrous outlet septum is malaligned in a postero-caudal fashion with respect to the trabecular muscular septum such that there is usually obstruction to the left ventricular outflow tract.		Doubly committed subarterial ventricular septal defect with muscular posterior inferior rim and posterior malalignment of outlet septum; Doubly committed juxta-arterial ventricular septal defect with muscular posterior inferior rim “coarctation type”	

Table 2. (*Continued*)

220	204	07.12.06	Doubly committed juxta-arterial ventricular septal defect with posteriorly malaligned fibrous outlet septum and perimembranous extension	A congenital cardiac malformation in which there is an outlet ventricular septal defect with perimembranous extension, bordered superiorly by the area of fibrous continuity or a fibrous outlet septum between the aortic and pulmonary valves, and the fibrous outlet septum is malaligned in a postero-caudal fashion with respect to the trabecular muscular septum such that there is usually obstruction to the left ventricular outflow tract.		Doubly committed subarterial ventricular septal defect with perimembranous extension and posterior malalignment of outlet septum; Doubly committed juxta-arterial ventricular septal defect with perimembranous extension "coarctation type"; Doubly committed juxta-arterial ventricular septal defect with posteriorly malaligned fibrous outlet septum and fibrous postero-inferior rim (perimembranous)
221	205	07.15.01	Ventricular septal defect haemodynamically insignificant	A congenital cardiac malformation in which there is one or more small, clinically insignificant ventricular septal defect(s) in the absence of flow-related cardiac chamber dilation or abnormal elevation of pulmonary arterial pressure.	Though restrictive ventricular septal defect is listed as a synonym of haemodynamically insignificant VSD, it should be recognized that some pressure restrictive ventricular septal defects will lead to flow-related chamber dilation, and thus would be haemodynamically significant. In such instances, the term haemodynamically insignificant ventricular septal defect should not be coded.	Maladie de Roger; Restrictive ventricular septal defect; Hemodynamically insignificant ventricular septal defect
222	206	07.15.04	Multiple ventricular septal defects	A congenital cardiac malformation in which there are multiple ventricular septal defects, which could be of any type.	For multiply fenestrated trabecular septums one should use the term "Multiple trabecular muscular ventricular septal defects".	
223	207	01.01.22	Functionally univentricular heart	The term "functionally univentricular heart" describes a spectrum of congenital cardiac malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary circulation.	A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions in this category typically include double inlet right ventricle (DIRV), double inlet left ventricle (DILV), tricuspid atresia, mitral atresia, and hypoplastic left heart syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include complex forms of atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations. Specific diagnostic codes should be used whenever possible, and not the term "functionally univentricular heart".	Univentricular cardiopathy; Single ventricle; Univentricular heart

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Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
224	208	01.01.14	Double inlet atrioventricular connection	A congenital cardiovascular malformation with a univentricular atrioventricular connection wherein both atria connect to one ventricle either via two separate atrioventricular valves or a common atrioventricular valve, such that all or nearly all of the total atrioventricular junctional (annular) area is committed to one ventricular chamber.		Double inlet ventricle	
225	209	01.04.04	Double inlet left ventricle	A congenital cardiovascular malformation with a univentricular atrioventricular connection wherein both atria connect to a morphologically left ventricle either via two separate atrioventricular valves or a common atrioventricular valve, such that all or nearly all of the total atrioventricular junctional (annular) area is committed to the left ventricular chamber.			DILV
226	210	01.04.03	Double inlet right ventricle	A congenital cardiovascular malformation with a univentricular atrioventricular connection wherein both atria connect to a morphologically right ventricle either via two separate atrioventricular valves or a common atrioventricular valve, such that all or nearly all of the total atrioventricular junctional (annular) area is committed to the right ventricular chamber.			DIRV
227	211	01.04.05	Double inlet to solitary ventricle of indeterminate morphology	A congenital cardiovascular malformation with a univentricular atrioventricular connection wherein both atria connect to a solitary and morphologically indeterminate ventricle either via two separate atrioventricular valves or a common atrioventricular valve.			
228	212	06.01.01	Tricuspid atresia	A congenital cardiovascular malformation with absence of the tricuspid valvar annulus (connection/junction) or an imperforate tricuspid valve.			

Table 2. (*Continued*)

229	213	06.01.26	Tricuspid atresia with absent atrioventricular connection	A congenital cardiovascular malformation with absence of the tricuspid valve and its associated annulus (connection/junction).	Tricuspid atresia with absent valvar annulus	
230	214	06.01.02	Tricuspid atresia with imperforate tricuspid valve	A congenital cardiovascular malformation with an imperforate tricuspid valve (the tricuspid valve and associated atrioventricular connection/junction are present but the valve is imperforate).		
231	215	06.02.01	Mitral atresia	A congenital cardiovascular malformation with absence of the mitral valvar annulus (connection/junction) or an imperforate mitral valve.		
232	216	06.02.26	Mitral atresia with absent atrioventricular connection	A congenital cardiovascular malformation with absence of the mitral valve and its associated annulus (connection/junction).	Mitral atresia with absent valvar annulus	
233	217	06.02.02	Mitral atresia with imperforate mitral valve	A congenital cardiovascular malformation with an imperforate mitral valve (the mitral valve and associated atrioventricular connection/junction are present but the valve is imperforate).		
234	218	01.01.09	Hypoplastic left heart syndrome	A spectrum of congenital cardiovascular malformations with normally aligned great arteries without a common atrioventricular junction, characterized by underdevelopment of the left heart with significant hypoplasia of the left ventricle including atresia, stenosis, or hypoplasia of the aortic or mitral valve, or both valves, and hypoplasia of the ascending aorta and aortic arch.	Hypoplasia of the left heart; hypoplasia of the left heart; HLHS; hlh - hypoplastic left heart syndrome; left heart hypoplasia syndrome	HLHS
235	219	09.04.29	Congenital anomaly of a ventriculo-arterial valve or adjacent regions	A congenital cardiovascular malformation of a ventriculo-arterial valve or its immediate subvalvar and supravalvar regions. This does not include anomalies of the truncal valve which are classified along with codes related to the common arterial trunk.		

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Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
236	220	09.05.29	Congenital anomaly of pulmonary valve	A congenital malformation of the heart where the pulmonary valve is abnormal.			
237	221	09.05.04	Congenital pulmonary valvar stenosis	A congenital cardiovascular malformation of the pulmonary valve in which there is narrowing or stricture causing obstruction to flow	Congenital pulmonary valvar stenosis ranges from critical neonatal pulmonic valve stenosis with hypoplasia of the right ventricle to valvar pulmonary stenosis in the infant, child, or adult.	Congenital pulmonary stenosis; Congenital pulmonary valve stricture; Congenital stenosis of pulmonary valve; Congenital pulmonary valve stenosis	
238	222	09.05.05	Pulmonary annular hypoplasia	A congenital cardiovascular malformation of the pulmonary valve in which its 'annulus' is hypoplastic (incomplete development or underdevelopment so that it is abnormally small [below the lower limit of normal adjusted for body size]).			
239	223	09.05.22	Congenital pulmonary regurgitation	A congenital cardiovascular malformation of the pulmonary valve allowing backward flow into the ventricle	Congenital pulmonary valve regurgitation may be due to primary annular dilation, prolapse, and leaflet underdevelopment.	Congenital pulmonary insufficiency; Congenital pulmonary incompetence; Congenital insufficiency of pulmonary valve	
240	224	09.05.24	Dysplasia of pulmonary valve	A congenital cardiovascular malformation where the pulmonary valve leaflets are markedly thickened with restricted mobility, characterized by the presence of myxomatous tissue.		Pulmonary valve dysplasia; Dysplastic pulmonary valve; Pulmonary valve cusp dysplasia	
241	225	09.05.32	Bicuspid pulmonary valve	A congenital cardiovascular malformation where the pulmonary valve has only two leaflets (cusps) and includes absent or fused commissure.		Bicommisural pulmonary valve	

Table 2. (*Continued*)

242	226	07.05.32	Congenital subpulmonary stenosis	A congenital cardiovascular malformation associated with narrowing within the outflow tract supporting the pulmonary valve.	Subvalvar (infundibular) pulmonary stenosis is a narrowing of the outflow tract of the ventricle immediately below the pulmonic valve. This term should preferably be used in the setting of abnormal ventriculo-arterial connections, such as double outlet ventricle. Although subpulmonary obstruction is a type of right ventricular outflow tract obstruction if the ventriculo-arterial connections are normal, in this setting 'Congenital right ventricular outflow tract obstruction' should be used. Subvalvar pulmonary stenosis is also a type of left ventricular outflow tract obstruction in the setting of discordant ventriculo-arterial connections; this term should be used when obstruction is only apparent immediately below the pulmonary valve, otherwise the term 'Congenital left ventricular outflow tract obstruction' should be used.	Infundibular pulmonary stenosis	
243	227	09.07.15	Congenital supravalvar pulmonary stenosis	A congenital cardiovascular malformation associated with narrowing at the level of the pulmonary sinotubular junction.			
244	228	09.05.16	Congenital pulmonary atresia	A congenital cardiovascular malformation in which there is no opening between any ventricle and the pulmonary arterial tree.	For "pulmonary atresia with VSD", please see the section under "Tetralogy of Fallot".	Pulmonary atresia.	
245	Q^{+}	09.05.12	Congenital pulmonary valvar atresia	A congenital cardiac malformation in which the pulmonary valve leaflet structure is present but imperforate.	This term is limited to describe hearts in which the pulmonary subvalvar or supravalvar regions are not atretic.	Congenital pulmonary valve atresia; Imperforate pulmonary valve; Pulmonary valvar atresia; Atretic pulmonary valve; Pulmonary valvular atresia.	
246	229	01.01.07	Pulmonary atresia with intact ventricular septum	A congenital cardiovascular malformation in which there are normally aligned great arteries, no opening between the morphologically right ventricle and the pulmonary trunk, and no ventricular level communication.	Pulmonary atresia with intact ventricular septum is a duct-dependent congenital malformation that forms a spectrum of lesions including atresia of the pulmonary valve, a varying degree of right ventricle and tricuspid valve hypoplasia, and anomalies of the coronary circulation. A right ventricular dependent coronary artery circulation is present when coronary artery fistulas are associated with a proximal coronary artery stenosis. Associated Ebstein anomaly of the tricuspid valve can be present.		PA/IVS

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Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
247	230	09.15.19	Congenital anomaly of aortic valve	A congenital cardiovascular malformation where the aortic valve is abnormal			
248	231	09.15.01	Congenital aortic valvar stenosis	A congenital cardiovascular malformation of the aortic valve in which there is narrowing or stricture (obstruction to flow). 'Congenital aortic valvar stenosis' arises most commonly as a result of partial or complete fusion of one or more commissures, or is due to dysplasia of one or more aortic cusps. These congenital malformations of the aortic valve may not be initially obstructive but may become stenotic later in life due to leaflet thickening, poor relative growth and-or calcification. It is not until the congenitally malformed aortic valve is or becomes stenotic that this term should be used.		Congenital valvar aortic stenosis; Congenital valvular aortic stenosis; Congenital aortic valve stricture; Congenital stenosis of aortic valve; Congenital aortic valve stenosis	AS
249	232	09.15.07	Congenital aortic regurgitation	A congenital cardiovascular malformation of the aortic valve allowing backward flow into the ventricle.	Congenital aortic regurgitation/ insufficiency is rare as an isolated entity. Aortic insufficiency is more commonly seen with other associated cardiovascular anomalies.	Congenital aortic incompetence; Congenital aortic insufficiency; AR - Congenital aortic regurgitation; Congenital aorta valve insufficiency; Congenital aortic regurgitation; Congenital insufficiency of aortic valve	Congenital AI; Congenital AR
250	233	09.15.22	Bicuspid aortic valve	A congenital cardiovascular malformation where the aortic valve has two commissures and two separate leaflets (cusps) because of fusion or absence of one of the commissures		Bicommisural aortic valve	BAV
251	234	09.15.21	Unicuspid aortic valve	A congenital cardiovascular malformation in which the aortic valve has a single commissure and a single or functionally single leaflet (cusp)		Unicommissural aortic valve	
252	235***	09.15.30	Aortic valvar prolapse	A congenital cardiovascular malformation of the aortic valve in which part or all of one or more of the aortic valve leaflets is on the ventricular side of the plane of the inferior aspect of the attachments of the aortic valve leaflets.		Aortic valve prolapse	

Table 2. (*Continued*)

253	236	09.15.06	Aortic valvar atresia	A congenital cardiovascular malformation in which there is no orifice of the aortic valve.	Aortic valve atresia will most often be coded under the hypoplastic left heart syndrome/complex diagnostic codes since it most often occurs as part of a spectrum of cardiovascular malformations. However, there is a small subset of patients with aortic valve atresia who have a well developed left ventricle and mitral valve and a large ventricular septal defect (nonrestrictive or restrictive).	Aortic valve atresia; Atresia of the aortic valve; Aorta valvular atresia; Aortic valvular atresia; Congenital aortic: atresia; Congenital atresia of aortic valve; Imperforate aortic valve	AA
254	237	09.15.17	Aortic annular hypoplasia	A congenital cardiovascular malformation of the aortic valve in which its 'annulus' is hypoplastic (incomplete development or underdevelopment so that it is abnormally small [below the lower limit of normal adjusted for body size]).		Hypoplasia of the aortic annulus	
255	238	09.15.09	Dysplasia of aortic valve	A congenital cardiovascular malformation where the aortic valve leaflets are markedly thickened with restricted mobility, characterized by the presence of myxomatous tissue.		Aortic valve dysplasia; Aortic valvar dysplasia	
256	239	07.09.50	Congenital subaortic stenosis	A congenital cardiovascular malformation associated with narrowing within the outflow tract supporting the aortic valve.	This term should be used for congenital subaortic stenosis due to lesions such as deviation of the outlet septum (seen in coarctation of the aorta and interrupted aortic arch), or a restrictive ventricular septal defect (bulboventricular foramen) in single ventricle complexes. This term should not be used for subaortic stenosis due to a fibromuscular shelf or tunnel, as specific terms exist for these lesions. For subaortic stenosis due to an atrioventricular valve in the setting of normal ventriculo-arterial connections, the term 'Congenital left ventricular outflow tract obstruction due to an atrioventricular valve' should be used.	Congenital subvalvular aortic stenosis; Subvalvular aortic stenosis, Congenital; Congenital subvalvar aortic stenosis	
257	240	07.09.03	Subaortic stenosis due to fibromuscular shelf	A cardiovascular malformation in which there is subaortic stenosis due to a discrete fibrous and/or muscular ridge.		Discrete LVOTO	

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Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
258	241	07.09.16	Subaortic stenosis due to fibromuscular tunnel	A cardiovascular malformation in which there is a long-segment fibrous and/or muscular subaortic stenosis.		Diffuse LVOTO	
259	242	09.16.18	Congenital supravalvar aortic stenosis	A congenital cardiovascular malformation with narrowing of the aorta at the level of the sinotubular junction which may extend into the ascending aorta.	Congenital supravalvar aortic stenosis is described as three forms: an hourglass deformity, a fibrous membrane, and a diffuse narrowing of the ascending aorta. Supravalvar aortic stenosis may involve the coronary artery ostia, and the aortic leaflets may be tethered. The coronary arteries can become tortuous and dilated due to elevated pressures and early atherosclerosis may ensue.	Stenosis at or above the sinotubular junction; Ascending aorta stenosis; Ascending aorta stricture	
260	243	09.18.01	Aneurysm of aortic sinus of Valsalva	A congenital cardiovascular malformation in which there is dilation of one or more aortic sinus of Valsalva.	The sinus of Valsalva is defined as that portion of the aortic root between the aortic root annulus and the sinotubular junction. Sinus of Valsalva aneurysm most commonly originates from the right sinus, less commonly from the non-coronary sinus, and rarely from the left sinus (<5%). The aneurysm may rupture into an adjacent chamber or site (right atrium, right ventricle, left atrium, left ventricle, pulmonary artery, pericardium) and in this case should be coded specifically ('Ruptured aortic sinus of Valsalva aneurysm'). This is to be distinguished from aortic root dilation associated with connective tissue disorders and aortopathies.	Aortic sinus of valsalva aneurysm	
261	244	09.17.01	Aortoventricular tunnel	A congenital cardiovascular malformation in which there is a paravalvar communication between the aorta and a ventricle.		Aortico-ventricular tunnel; Ventriculo-aortic tunnel; Aorto-ventricular tunnel	
262	R^{+}	09.17.02	Aorto-left ventricular tunnel	A congenital cardiovascular malformation in which there is a paravalvar communication between the ascending aorta and the left ventricle.			

Table 2. (*Continued*)

263	S^+	09.17.04	Aorto-right ventricular tunnel	A congenital cardiovascular malformation in which there is a paravalvar communication between the ascending aorta and the right ventricle		
264	245	09.04.28	Congenital anomaly of great arteries including arterial duct	A congenital cardiovascular malformation of the great arteries (aorta, pulmonary trunk [main pulmonary artery], branch pulmonary arteries) or the arterial duct (ductus arteriosus).	This term excludes the truncal root in "Common arterial trunk" (truncus arteriosus).	
265	246	09.04.07	Congenital aortopulmonary window	A congenital cardiovascular malformation in which there is side-to-side continuity of the lumens of the ascending aorta and pulmonary trunk in association with separate aortic and pulmonary valves or their atretic remnants.	Side-to-side continuity of the lumens of the aorta and pulmonary arterial tree, which is distinguished from common arterial trunk (truncus arteriosus) by the presence of two arterial valves or their atretic remnants, and involvement of the pulmonary trunk (main pulmonary artery).	Aortopulmonary window; Aortic septal defect; Aortopulmonary septal defect; aorticopulmonary window; aorticopulmonary fenestration; aorticopulmonary septal defect
266	247	09.07.16	Congenital anomaly of pulmonary arterial tree	A congenital cardiovascular malformation of the pulmonary trunk (main pulmonary artery) and/or branch pulmonary arteries (right, left, and ramifications).		
267	248	09.10.36	Congenital dilation of pulmonary arterial tree	A congenital cardiovascular malformation in which there is an enlargement of the luminal diameter of the pulmonary trunk (main pulmonary artery) and/or branch pulmonary arteries (above the upper limit of normal adjusted for body size).	Includes idiopathic isolated PA dilation	
268	249	09.07.19	Congenital pulmonary trunk anomaly	A congenital cardiovascular malformation of the pulmonary arterial trunk (main pulmonary artery).		Congenital main pulmonary artery anomaly
269	250	09.07.20	Congenital pulmonary trunk hypoplasia	A congenital cardiovascular malformation of the pulmonary trunk (main pulmonary artery) in which there is incomplete development or underdevelopment resulting in diffuse luminal narrowing (below the lower limit of normal adjusted for body size).	The stenosis or hypoplasia may be isolated or associated with other cardiovascular lesions. Since the narrowing is distal to the pulmonary valve, it may also be known as supravalvar pulmonary stenosis.	Main pulmonary artery hypoplasia

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Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
270	251	09.07.05	Absent or atretic pulmonary trunk	A congenital cardiovascular malformation where the pulmonary trunk (main pulmonary artery) is not present or has luminal occlusion, excluding common arterial trunk.		Pulmonary artery atresia; Atretic pulmonary trunk; Pulmonary trunk absent; Absent main pulmonary artery; Atretic main pulmonary artery; Absent or atretic main pulmonary artery	
271	252	09.10.41	Congenital pulmonary arterial branch anomaly	A congenital cardiovascular malformation of a pulmonary arterial branch.		Congenital abnormality of pulmonary artery	
272	253	09.10.27	Congenital pulmonary arterial branch stenosis				
273	254	09.10.28	Congenital right pulmonary arterial stenosis	A congenital cardiovascular malformation in which there is discrete narrowing of the luminal diameter of one or more segments of the right pulmonary artery (below the lower limit of normal adjusted for body size).			
274	255	09.10.29	Congenital left pulmonary arterial stenosis	A congenital cardiovascular malformation in which there is discrete narrowing of the luminal diameter of one or more segments of the left pulmonary artery (below the lower limit of normal adjusted for body size).			
275	256	09.10.71	Congenital pulmonary arterial branch hypoplasia	A congenital cardiovascular malformation in which there is diffuse luminal narrowing of a pulmonary arterial branch (below the lower limit of normal adjusted for body size).		Hypoplasia of pulmonary artery; congenital hypoplasia of pulmonary artery; pulmonary artery hypoplasia	
276	257	09.10.72	Congenital right pulmonary arterial hypoplasia	A congenital cardiovascular malformation in which there is diffuse luminal narrowing of the right pulmonary artery (below the lower limit of normal adjusted for body size).			
277	258	09.10.73	Congenital left pulmonary arterial hypoplasia	A congenital cardiovascular malformation in which there is diffuse luminal narrowing of the left pulmonary artery (below the lower limit of normal adjusted for body size).			

Table 2. (*Continued*)

278	T+	09.10.21	Absent or atretic right or left pulmonary artery	A congenital cardiovascular malformation in which the right and/or left pulmonary artery is not present or has luminal occlusion.		Atretic right or left pulmonary artery; Absent right or left pulmonary artery; Agenesis of pulmonary artery; Pulmonary arterial agenesis	
279	259	09.10.75	Absent or atretic right pulmonary artery	A congenital cardiovascular malformation in which the right pulmonary artery is not present or has luminal occlusion.		Atretic right pulmonary artery; Absent right pulmonary artery	
280	260	09.10.77	Absent or atretic left pulmonary artery	A congenital cardiovascular malformation in which the left pulmonary artery is not present or has luminal occlusion.		Atretic left pulmonary artery; Absent left pulmonary artery	
281	261	09.10.37	Congenital central pulmonary arterial stenosis or hypoplasia proximal to hilar bifurcation	A congenital cardiovascular malformation of a pulmonary artery, proximal to its first branch, in which there is luminal narrowing (below the lower limit of normal adjusted for body size).	The stenosis or hypoplasia may be isolated or associated with other cardiovascular lesions. Coarctation of the pulmonary artery is related to abnormal extension of the arterial duct (ductus arteriosus) into a pulmonary arterial branch, more frequently the left branch. This is to be distinguished from narrowing or hypoplasia of the pulmonary trunk (main pulmonary artery).	Central pulmonary arterial stenosis; Central pulmonary stenosis; Proximal pulmonary arterial stenosis; Proximal pulmonary stenosis	
282	262	09.10.38	Congenital peripheral pulmonary arterial stenosis or hypoplasia at or beyond hilar bifurcation	A congenital cardiovascular malformation of a pulmonary artery, distal to its first branch, in which there is luminal narrowing (below the lower limit of normal adjusted for body size).	Peripheral pulmonary artery narrowing or hypoplasia at or beyond the hilar bifurcation. The stenosis or hypoplasia may be isolated or associated with other cardiovascular lesions.	Peripheral pulmonary stenosis; Distal peripheral pulmonary stenosis; Peripheral pulmonary arterial stenosis; Distal peripheral pulmonary arterial stenosis	
283	263	09.10.30	Congenitally discontinuous, non-confluent right and left pulmonary arteries	A congenital cardiovascular malformation in which there is absence of luminal continuity between the right and left branch pulmonary arteries.	The discontinuous branch pulmonary artery is typically supplied by a patent arterial duct (ductus arteriosus) or an aortopulmonary collateral.		
284	264	09.09.08	Pulmonary artery origin from ascending aorta	A congenital cardiovascular malformation in which one branch pulmonary artery arises from the ascending aorta and the other branch pulmonary artery arises from the pulmonary trunk (main pulmonary artery).	One pulmonary artery arises from the ascending aorta and the other pulmonary artery arises from the right ventricle. This does include origin of the right or left pulmonary artery from the innominate (brachiocephalic) artery or the aortic arch via a patent arterial duct (ductus arteriosus) or collateral artery.		Hemitruncus

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Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
285	265	09.09.03	Right pulmonary artery from ascending aorta	A congenital cardiovascular malformation in which the right pulmonary artery arises from the ascending aorta and the left pulmonary artery arises from the pulmonary trunk (main pulmonary artery).			Hemitruncus
286	266	09.09.05	Left pulmonary artery from ascending aorta	A congenital cardiovascular malformation in which the left pulmonary artery arises from the ascending aorta and the right pulmonary artery arises from the pulmonary trunk (main pulmonary artery).			Hemitruncus
287	267	09.09.11	Pulmonary artery from arterial duct	A congenital cardiovascular malformation in which the pulmonary arteries are non-confluent and one or both arise from an arterial duct (ductus arteriosus).	When both arise from AD then code under discontinuous	Pulmonary artery from ductus arteriosus	
288	268	09.09.02	Right pulmonary artery from arterial duct	A congenital cardiovascular malformation in which the pulmonary arteries are non-confluent and the right pulmonary artery arises from an arterial duct (ductus arteriosus).		Right pulmonary artery from ductus arteriosus	
289	269	09.09.04	Left pulmonary artery from arterial duct	A congenital cardiovascular malformation in which the pulmonary arteries are non-confluent and the left pulmonary artery arises from an arterial duct (ductus arteriosus).		Left pulmonary artery from ductus arteriosus	
290	270	07.09.34	Congenital anomaly of aorta or its branches	A congenital cardiovascular malformation of the aorta and/or its branches.			
291	271	09.16.06	Congenital anomaly of ascending aorta	A congenital cardiovascular malformation of the aorta between the sinotubular junction and the origin of its first branch.			
292	272	09.16.02	Hypoplasia of ascending aortic	A congenital cardiovascular malformation in which the luminal diameter of the aorta between its sinotubular junction and the origin of the innominate (brachiocephalic) artery is narrowed (below the lower limit of normal adjusted for body size).		Ascending aortic hypoplasia	

Table 2. (*Continued*)

293	273	09.16.19	Congenital ascending aortic aneurysm or dilation	A congenital cardiovascular malformation in which the luminal diameter of the aorta between its sinotubular junction and the origin of the innominate (brachiocephalic) artery is dilated (above the upper limit of normal adjusted for body size).			
294	274	09.28.10	Congenital anomaly of aortic arch	A congenital cardiovascular malformation of the aorta between the origin of the innominate artery and the insertion of the arterial duct (ductus arteriosus).			
295	275	09.29.11	Hypoplasia of aortic arch	A congenital cardiovascular malformation in which there is diffuse luminal narrowing of the aortic arch (below the lower limit of normal adjusted for body size).	Hypoplasia of the aortic arch is hypoplasia of the proximal or distal transverse arch or the aortic isthmus. The isthmus (arch between the left subclavian and insertion of the patent ductus arteriosus/ligamentum arteriosum) is hypoplastic if its diameter is less than 40% of the diameter of the ascending aorta. The proximal transverse arch (arch between the innominate and left carotid arteries) and distal transverse arch (arch between the left carotid and left subclavian arteries) are hypoplastic if their diameters are less than 60% and 50%, respectively, of the diameter of the ascending aorta.	Hypoplastic aortic arch; Arch hypoplasia; Aortic hypoplasia; Aortic arch hypoplasia	
296	277	09.29.31	Interrupted aortic arch	A congenital cardiovascular malformation in which there is an absence of luminal continuity between the ascending and descending aorta.	This term includes luminal atresia with discontinuity between the aortic segments and also luminal atresia with fibrous continuity between the aortic segments. Interrupted aortic arch is defined as the loss of luminal continuity between the ascending and descending aorta. In most cases, blood flow to the descending thoracic aorta is through a patent arterial duct, and there is a large ventricular septal defect. Arch interruption is further defined by site of interruption. In type A, interruption is distal to the left subclavian artery; in type B, interruption is between the left carotid and left subclavian arteries; and in type C, interruption occurs between the innominate and left carotid arteries.	Aortic arch interruption, aortic interruption, interruption of the aortic arch, aortic atresia	IAA

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Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
297	278	09.29.32	Interrupted aortic arch distal to subclavian artery, type A	A congenital cardiovascular malformation of the aorta in which there is an absence of luminal continuity distal to the subclavian artery ipsilateral to the arch.		Aortic arch interruption Type A; Aortic interruption Type A; Interruption of the aortic arch Type A	IAA Type A
298	279	09.29.33	Interrupted aortic arch between subclavian and common carotid arteries, type B	A congenital cardiovascular malformation of the aorta in which there is an absence of luminal continuity of the aorta between the carotid and subclavian arteries.		Aortic arch interruption Type B; Aortic interruption Type B; Interruption of the aortic arch Type B	IAA Type B
299	280	09.29.34	Interrupted aortic arch between carotid arteries, type C	A congenital cardiovascular malformation of the aorta in which there is an absence of luminal continuity of the aorta between the carotid arteries.		Aortic arch interruption Type C; Aortic interruption Type C; Interruption of the aortic arch Type C	IAA Type C
300	281	09.28.15	Right aortic arch	A congenital cardiovascular malformation of the great vessels in which the aortic arch crosses to the right of the trachea.		Right arch; Right-sided arch; Right-sided aortic arch; Persistent right arch; Persistent right-sided arch; Persistent right-sided aortic arch	
301	282	09.28.22	Left aortic arch	A congenital cardiovascular finding of the great vessels in which the aortic arch crosses to the left of the trachea.	To be coded only when this represents an abnormal finding, such as in situs inversus.	Left arch; Left-sided arch; Left-sided aortic arch	
302	283	09.28.06	Cervical aortic arch	A congenital cardiovascular malformation in which the aortic arch is located superior to the clavicle, and is most commonly right-sided.		Cervical arch	
303	X^{+}	09.30.22	Aortic diverticulum of Kommerell	A congenital cardiovascular malformation consisting of an aneurysmally dilated proximal portion of an aberrant subclavian artery or aberrant innominate (brachiocephalic) artery as it arises from the descending aorta.			

Table 2. (*Continued*)

304	Y+	09.28.08	Persistent fifth aortic arch	A congenital cardiovascular malformation in which there is an accessory artery originating from the ascending aorta proximal to the ostium of the innominate (brachiocephalic) artery which connects to the descending aorta or near the confluence of the right and left pulmonary arteries.	When the arterial connection is between the ascending and descending aorta, it is often associated with coarctation of the aorta. This malformation may be distinguished from a double aortic arch by the lack of arch vessels arising from it and by establishing that the accessory vessel and the aortic arch lie on the same side of the tracheo-oesophageal axis. Although this malformation has been termed a "persistent fifth aortic arch", the use of this term is in dispute.	
305	276	09.29.01	Coarctation of aorta	A congenital cardiovascular malformation in which there is a discrete luminal narrowing of the junction between the aortic arch and the descending aorta.	Coarctation of the aorta generally indicates a narrowing of the descending thoracic aorta just distal to the left subclavian artery. However, the term may also be accurately used to refer to a region of narrowing anywhere in the thoracic or abdominal aorta.	Aortic coarctation
306	U+	09.29.02	Preductal coarctation of aorta	A congenital cardiovascular malformation in which there is narrowing of the aortic lumen proximal to the insertion of the arterial duct (ductus arteriosus) or ligament (ligamentum arteriosum).		
307	V+	09.29.04	Postductal coarctation of aorta	A congenital cardiovascular malformation in which there is narrowing of the aortic lumen distal to the insertion of the arterial duct (ductus arteriosus) or ligament (ligamentum arteriosum).		
308	W+	09.29.03	Juxtaductal (paraductal) coarctation of aorta	A congenital cardiovascular malformation in which there is narrowing of the aortic lumen opposite the level of insertion of the arterial duct (ductus arteriosus) or ligament (ligamentum arteriosum).		
309	284	09.30.17	Congenital anomaly of aortic arch branch	A congenital cardiovascular malformation of one or more branches of the aortic arch (innominate, carotid, or subclavian arteries).		

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Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
310	285	09.30.02	Aberrant origin of right subclavian artery	A congenital cardiovascular malformation in which the right subclavian artery arises distal to the left subclavian artery in the setting of a left aortic arch.	Dysphagia lusoria (or Bayford-Autenrieth dysphagia) is an abnormal condition characterized by difficulty in swallowing caused by an aberrant right subclavian artery. It was discovered by David Bayford in 1761 and first reported in a paper by the same in 1787	Aberrant right subclavian artery; Arteria lusoria; Dysphagia lusoria; Aberrant right subclavian artery syndrome	ARSA, ARSCA
311	286	09.30.04	Aberrant origin of left subclavian artery	A congenital cardiovascular malformation in which the left subclavian artery arises distal to the right subclavian artery in the setting of a right arch.		Aberrant left subclavian artery; Retroesophageal left subclavian artery	ALSA, ALSCA, RELSCA
312	287	09.30.16	Isolation of an aortic arch branch	A congenital cardiovascular malformation in which the only connection to a brachiocephalic vessel is via an arterial duct (ductus arteriosus) or its ligament.			
313	Z^{+}	09.30.11	Isolation of innominate artery	A congenital cardiovascular malformation in which the innominate (brachiocephalic) artery has no direct connection to the aorta.		Isolation of brachiocephalic artery; Isolation of brachiocephalic trunk	
314	AA^{+}	09.30.14	Isolation of left subclavian artery	A congenital cardiovascular malformation in which the left subclavian artery has no direct connection to the aorta.			
315	AB^{+}	09.30.15	Isolation of right subclavian artery	A congenital cardiovascular malformation in which the right subclavian artery has no direct connection to the aorta.			
316	AC^{+}	09.30.12	Isolation of left common carotid artery	A congenital cardiovascular malformation in which the left common carotid artery has no direct connection to the aorta.			
317	AD^{+}	09.30.13	Isolation of right common carotid artery	A congenital cardiovascular malformation in which in which the right common carotid artery has no direct connection to the aorta.			

Table 2. (*Continued*)

318	AE+	09.30.34	Aberrant origin of innominate artery	A congenital cardiovascular malformation in which an innominate (brachiocephalic) artery arises from an abnormally distal position on the aortic arch, or from a diverticulum of Kommerell, or from a pulmonary artery.		Aberrant origin of brachiocephalic artery; Aberrant origin of brachiocephalic trunk	
319	AF+	09.30.31	Common origin of the innominate artery and left common carotid artery	A congenital cardiovascular malformation in which the innominate (brachiocephalic) artery and left common carotid artery arise from a single aortic orifice in the setting of a left aortic arch.	This term as well as "Left common carotid arising from innominate artery" have been used synonymously (but incorrectly) with the term "Bovine aortic arch", despite the fact that neither resemble a true bovine arch.		
320	AG+	09.30.28	Separate origins of internal and external carotid arteries	A congenital cardiovascular malformation in which there is an absence of a common carotid artery and the internal and external carotid arteries connect separately to the aorta.			
321	288	09.28.47	Congenital anomaly of descending thoracic or abdominal aorta	A congenital cardiovascular malformation of the aorta distal to the aortic arch.		Congenital anomaly of thoracoabdominal aorta	
322	289	09.29.44	Descending thoracic or abdominal aortic coarctation	A congenital cardiovascular malformation in which there is discrete luminal narrowing of the descending thoracic or abdominal aorta.		Middle aortic syndrome	MAS
323	AH+	09.29.05	Coarctation of the descending thoracic aorta	A congenital cardiovascular malformation in which there is discrete luminal narrowing of the descending thoracic aorta.			
324	AI+	09.29.06	Coarctation of the abdominal aorta	A congenital cardiovascular malformation in which there is discrete luminal narrowing of the abdominal aorta.			
325	290	09.31.40	Tracheo-oesophageal compressive syndrome	A congenital cardiovascular malformation which causes compression of the trachea and/or the oesophagus.			

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Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
326	291	09.30.23	Innominate artery compression syndrome	A congenital cardiovascular malformation in which there is anterior compression of the trachea by the innominate artery.	This syndrome is a true compression of the trachea by the abnormally positioned innominate artery. The innominate artery can appear to "compress" the trachea in the presence of tracheomalacia but, in the absence of an abnormal origin and course of the innominate artery, this apparent "compression" most likely is a consequence of the tracheomalacia itself rather than actual compression by the innominate artery.	Innominate artery compressive syndrome; Brachiocephalic artery compression syndrome; Brachiocephalic artery compressive syndrome; Innominate artery compression of the trachea; Brachiocephalic artery compression of the trachea; Aberrant innominate artery; Aberrant brachiocephalic artery; Anomalous origin of the innominate artery; Anomalous origin of the brachiocephalic artery	
327	AJ^{+}	09.30.27	Retro-oesophageal origin of aberrant innominate artery	A congenital cardiovascular malformation in which the innominate (brachiocephalic) artery passes from right-to-left or from left-to-right posterior to the oesophagus.			
328	292	09.31.00	Vascular Ring	A congenital cardiovascular malformation in which one or more of the following encircle the trachea and oesophagus: the aorta and its major branches, the pulmonary trunk and its major branches, and the arterial duct (ductus arteriosus) or their vascular remnant(s).	The term vascular ring refers to a group of congenital vascular anomalies that encircle and compress the oesophagus and/or trachea. The compression may be from a complete anatomic ring (double aortic arch or right aortic arch with a left ligamentum) or from a compressive effect of an aberrant vessel (innominate artery compression syndrome).		
329	293	09.28.09	Double aortic arch	A congenital cardiovascular malformation in which the right and left aortic arches (patent or atretic) encircle the trachea and oesophagus.		Encircling double aortic arch	DAA
330	294	09.31.35	Vascular ring of right aortic arch and left arterial duct or ligament	A congenital cardiovascular malformation in which continuity of the right aortic arch, pulmonary trunk and left arterial duct (or ligament) encircles the trachea and oesophagus.	This diagnosis may or may not include the presence of a diverticulum of Kommerell.		
331	295	09.31.34	Vascular ring of left aortic arch and right arterial duct or ligament	A congenital cardiovascular malformation in which continuity of the left aortic arch, pulmonary trunk and right arterial duct (or ligament) encircles the trachea and oesophagus.	This diagnosis may or may not include the presence of a diverticulum of Kommerell.		

Table 2. (*Continued*)

332	296	09.09.06	Anomalous origin of left pulmonary artery from right pulmonary artery	A congenital cardiovascular malformation in which the left pulmonary artery originates from the right pulmonary artery and passes between the trachea and oesophagus, and is often associated with tracheobronchial anomalies such as tracheomalacia, stenosis, or complete tracheal rings.		Pulmonary arterial sling	
333	297	09.27.05	Congenital arterial duct anomaly	A congenital cardiovascular malformation of the arterial duct (ductus arteriosus) or its fibrous remnant (ligamentum arteriosum).	The described anomalies include an anomalous course, abnormal laterality or duplication, persistent patency or premature closure, and aneurysm formation.	Congenital ductus arteriosus anomaly	Congenital PDA anomaly
334	298	09.27.21	Patent arterial duct	A congenital cardiovascular finding in which the arterial duct (ductus arteriosus) is open beyond the normal age of spontaneous closure.	A patent arterial duct (ductus arteriosus) is a vascular arterial connection between the thoracic aorta and the pulmonary artery. Most commonly, a patent arterial duct has its origin from the descending thoracic aorta, just distal and opposite the origin of the left subclavian artery. The insertion of the ductus is most commonly into the very proximal left pulmonary artery at its junction with the main pulmonary artery. Origination and insertion sites can be variable, however.	Patent ductus arteriosus; Persistent ductus Botalli; Patent ductus Botalli; Open ductus arteriosus; Persistent ductus arteriosus; Ductus arteriosus nonclosure; Patent ductus arteriosus - persisting type; PDA - patent ductus arteriosus; Persistent ductus arteriosus (Botalli)	PDA; PAD
335	AK^{+}	09.27.03	Absent arterial duct	A congenital cardiovascular malformation in which the arterial duct (ductus arteriosus) or ligament (ligamentum arteriosum) is not present.		Absent ductus arteriosus; Ductus arteriosus agenesis	Absent PDA; Absent PAD
336	AL^{+}	09.27.04	Congenital aneurysm of arterial duct	A congenital cardiovascular malformation where there is fusiform or saccular dilatation of the arterial duct (ductus arteriosus).		Aneurysm of ductus arteriosus	Aneurysm of PDA; Aneurysm of PAD
337	AM^{+}	09.27.41	Anomalous origin of arterial duct	A congenital cardiovascular malformation in which there is an arterial duct (ductus arteriosus) that is not in its expected position between the aorta or arch vessels and the pulmonary arterial circulation.	While in the setting of a left aortic arch the arterial duct is expected to arise from the concavity of the aortic arch, in the case of a right aortic arch it can normally be expected to arise from the base of the innominate (brachiocephalic) artery.	Anomalous origin of ductus arteriosus	Anomalous origin of PDA; Anomalous origin of PAD

(*Continued*)

Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
338	AN^{+}	09.27.82	Anomalous origin of arterial ligament	A congenital cardiovascular malformation in which there is a remnant of the arterial duct (ductus arteriosus) that is not in its expected position between the aorta or arch vessels and the pulmonary arterial circulation.	While in the setting of a left aortic arch the arterial ligament is expected to be seen stretching from the concavity of the aortic arch to the pulmonary artery confluence, in the case of a right aortic arch one end of the ligament can normally be found at the base of the innominate (brachiocephalic) artery.		
339	AO^{+}	14.10.51	Fetal arterial duct narrowing-closure	A congenital cardiovascular malformation in which there is prenatal partial or complete closure of the arterial duct (ductus arteriosus).		Fetal ductus arteriosus narrowing-closure	Fetal PDA narrowing-closure; Fetal PAD narrowing-closure
340	299	09.08.18	Systemic-to-pulmonary collateral arteries	A congenital cardiovascular malformation in which the blood supply to the lungs is derived completely or in part from collateral vessels that arise from the aorta or its branches.	At least part of the pulmonary blood flow is derived from systemic-to-pulmonary collateral arteries that are highly variable in number, and that usually arise from the descending thoracic aorta, but uncommonly may originate from the aortic arch or the subclavian, carotid or even the coronary arteries. There may or may not be native pulmonary arteries which, if present, may be hypoplastic, and either confluent or non-confluent. This term is intended to exclude patients with systemic to pulmonary artery collaterals and "Tetralogy of Fallot with pulmonary atresia" since they are coded using the term "Tetralogy of Fallot with pulmonary atresia and systemic-to-pulmonary collateral arteries".	Major systemic-to-pulmonary collateral arteries; Major systemic-to-pulmonary collateral artery; Systemic-to-pulmonary collateral vessels; Systemic-to-pulmonary collateral flow; Aortopulmonary collateral arteries	APCAs, MAPCAs
341	300	09.46.03	Congenital anomaly of coronary artery	A congenital cardiovascular malformation of a coronary artery.	This includes absence of a coronary, anomalous origin or course, dilation or stenosis, and fistulas. Congenital anomalies of the coronary venous system should not be included here but rather under 'Congenital anomaly of mediastinal systemic vein'.	Congenital malformations of coronary vessels; malformation of coronary vessels	

Table 2. (*Continued*)

342	301	09.41.01	Anomalous origin of coronary artery from pulmonary arterial tree	A congenital cardiovascular malformation in which a coronary artery originates from the pulmonary trunk or one of its branches.	Although the most common of these malformations involves the left coronary artery arising from the pulmonary trunk (main pulmonary artery) rather than from the aorta, occasionally the right coronary artery, the circumflex, or both coronary arteries may arise from any of the central pulmonary arteries.		
343	302	09.41.03	Anomalous origin of left coronary artery from pulmonary artery	A congenital cardiovascular malformation in which the left coronary artery originates from the pulmonary trunk or one of its branches.		Anomalous origin of left coronary artery from pulmonary trunk (ALCAPT)	ALCAPA; ALCAPT
344	303	09.42.00	Anomalous aortic origin or course of coronary artery	A congenital cardiovascular malformation in which the origin and/or course of a coronary artery is abnormal.	This is where coronary "anomalies" in the presence of discordant ventriculo-arterial connections should be coded.		
345	304	09.42.21	Anomalous aortic origin of coronary artery with ventriculo-arterial concordance	A congenital cardiovascular malformation in the setting of ventriculo-arterial concordance in which a coronary artery arises from the aorta at a location other than its expected sinus.	Anomalous aortic origins of the coronary arteries include a spectrum of anatomic variations of the normal coronary artery origins. Coronary artery anomalies of aortic origin to be coded under this diagnostic field include: anomalies of take-off (high take-off), origin (sinus), branching, and number. An anomalous course of the coronary artery vessels is also significant, particularly those coronary arteries that arise or course between the great vessels. This term is used for patients with concordant ventriculo-arterial connections.	AAOCA; Anomalous aortic origin of coronary artery	AAOCA
346	305	09.46.26	Right coronary artery from left aortic sinus with ventriculo-arterial concordance	A congenital cardiovascular malformation in the setting of ventriculo-arterial concordance in which the right coronary artery arises from, or immediately above, the left sinus of Valsalva.	This term applies to patients with concordant ventriculo-arterial connections.		
347	306	09.46.21	Left coronary artery from right aortic sinus with ventriculo-arterial concordance	A congenital cardiovascular malformation in the setting of ventriculo-arterial concordance in which the left coronary artery arises from, or immediately above, the right sinus of Valsalva.	This term applies to patients with concordant ventriculo-arterial connections.		
348	307	09.43.04	Anterior descending from right coronary artery	A congenital cardiovascular malformation in which the left circumflex coronary artery arises		Anterior interventricular artery from right coronary artery across right ventricular outflow tract; Left	

(*Continued*)

Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
			across right ventricular outflow tract	normally and the anterior descending coronary artery arises from the proximal right coronary artery and courses across the right ventricular outflow tract.		anterior descending (LAD) from right coronary artery across right ventricular outflow tract	
349	308	09.43.05	Intramural proximal coronary arterial course	A congenital cardiovascular malformation in which the proximal coronary artery courses within and parallel to the wall of the aorta before it emerges to assume its epicardial course.		Intramural coronary artery	
350	309	09.43.13	Single coronary artery supplying all of heart	A congenital cardiovascular malformation in which a solitary coronary artery supplies the myocardium.			
351	310	09.43.12	Myocardial bridging of coronary artery	A congenital cardiovascular malformation in which a usually epicardial coronary arterial segment is located within the ventricular myocardium, making this segment susceptible to compression during systole.		Myocardial bridge	MB
352	311	09.44.05	Congenital coronary arterial orifice stenosis	A congenital cardiovascular malformation in which the orifice of a coronary artery is narrowed.		Congenital coronary ostial stenosis	
353	312	09.44.19	Congenital coronary arterial orifice atresia	A congenital cardiovascular malformation in which the orifice of a coronary artery is not patent.	This excludes single coronary artery anomalies	Congenital coronary ostial atresia	COSA
354	313	09.45.16	Congenital coronary arterial fistula	A congenital cardiovascular malformation in which a coronary artery communicates, through an anomalous channel, with a cardiac chamber or with any segment of the pulmonary circulation.	This communication may be simple and direct or may be tortuous and dilated. In order of frequency the involved coronary artery is the right, the left and, rarely, both coronary arteries. Occasionally multiple fistulas are present.	Coronary fistula	

Table 2. (*Continued*)

355	314	09.45.10	Congenital coronary arterial fistula to right ventricle	A congenital cardiovascular malformation in which a coronary artery communicates, through an anomalous channel, with the right ventricle.	
356	315	09.45.22	Congenital coronary arterial fistula to left ventricle	A congenital cardiovascular malformation in which a coronary artery communicates, through an anomalous channel, with the left ventricle.	
357	316	09.46.14	Congenital coronary arterial aneurysm	A congenital cardiovascular malformation in which there is one or more localized dilation(s) of a coronary vessel.	Coronary artery aneurysms are usually seen in two forms, saccular (shaped like a sack, with a change in caliber over a very short distance), and fusiform (shaped like a spindle, tapering towards each end), and both may be single or multiple.
358	AP^{+}	09.42.09	Accessory coronary artery	A congenital cardiac malformation in which there is an additional coronary artery that duplicates the blood supply of an existing and named coronary artery.	
359	AQ^{+}	09.46.44	Congenital absence of coronary artery	A congenital cardiac malformation in which a right, left main, interventricular, or circumflex coronary artery is not present in its anticipated epicardial course.	
360	AR^{+}	09.46.19	Coronary arterial hypoplasia	A congenital cardiovascular malformation in which one or more coronary arteries have an abnormally reduced length or lumen diameter.	
361	317	10.01.05	Congenital pericardial anomaly	A congenital malformation in which there is a structural and/or functional abnormality of the pericardium.	This term can include complete or partial absence of pericardium, pericardial cysts, antenatal pericardial effusion and congenital tumours of the serous pericardium.
362	AS^{+}	10.01.02	Complete agenesis of pericardium	A congenital cardiac malformation in which the fibroserous pericardium is completely absent.	

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Table 2. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	Definition	Commentary	Synonyms	Abbreviations
363	AT^{+}	10.01.01	Partial agenesis of pericardium	A congenital cardiac malformation in which the fibroserous pericardium is partly absent.			
364	AU^{+}	10.01.03	Pleuropericardial cyst	A congenital finding in which there is a closed sac typically found at the pleuropericardial angle, having a distinct membrane and division compared to the nearby tissue.	It may contain air, fluid, or semi-solid material. Congenital non-malignant pleuropericardial cysts include pericardial teratoma, cystic lymphangioma, bronchogenic, and celomic cysts.		
365	AV^{+}	10.03.53	Congenital cardiac tumour	A congenital malformation consisting of growth of abnormal tissue within the heart.			
366	318***	09.19.05	Pulmonary arteriovenous fistula	A congenital cardiovascular malformation in which there is an abnormal, direct connection between a pulmonary artery and pulmonary vein or left atrium without an intervening capillary bed.		Pulmonary arteriovenous malformation; Pulmonary arteriovenous aneurysm	
367	AW^{+}	02.02.03	Bifid apex of heart	A congenital cardiac finding in which there is a notch or cleft on the epicardial surface of the heart at the level of the distal interventricular groove that divides the apex in two, so that the apex of the left ventricle lies on one side of the cleft while the apex of the right ventricle lies on the other.	A bifid cardiac apex can be associated with congenital malformations but may also be found in otherwise normal hearts.		

Table 3. IPCCC ICD-11 Codes

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	ICD-11 Foundation entity number	ICD-11 MMS code or ICD-11 MMS crossmap	Parent	Sibling order	ICD-11 Congenital Cardiac term published in 2017[105] (terms in blue cells have been changed)	ICD-11 Congenital Cardiac term components published in 2017[105] and now moved to Synonyms
1	1	01.01.59	Structural developmental anomaly of heart or great vessels	2004408087	LA8Z	No parent - top of list	0	Structural developmental anomaly of heart and great vessels	
2	2	03.01.13	Congenital anomaly of position or spatial relationships of thoraco-abdominal organs	422322292	LA8Y	01.01.59	1	Congenital anomaly of position or spatial relationships of thoraco-abdominal organs	
3	3	02.01.09	Anomalous position-orientation of heart	731798335	LA80	03.01.13	1	Anomalous position-orientation of heart	
4	4**	02.01.03	Laevocardia	848076902	LA80.0	02.01.09	1	Laevocardia	
5	5	02.01.02	Dextrocardia	1472687600	LA80.1	02.01.09	2	Dextrocardia	
6	6	02.01.04	Mesocardia	1251061251	LA80.2	02.01.09	3	Mesocardia	
7	7	02.01.01	Extrathoracic heart	285576893	LA80.3	02.01.09	4	Extrathoracic heart	
8	8**	01.03.00	Usual atrial arrangement	1486434040		03.01.13	2	Usual atrial arrangement (atrial situs solitus)	Atrial situs solitus
9	9	01.03.06	Abnormal atrial arrangement	152930652	LA8Y	03.01.13	3	Abnormal atrial arrangement (abnormal atrial situs)	Abnormal atrial situs
10	10	01.03.01	Atrial situs inversus	1956091337	LA8Y	01.03.06	1	Atrial situs inversus	
11	11	01.03.02	Isomerism of right atrial appendages	967750556	LA8Y	01.03.06	2	Isomerism of right atrial appendages	
12	12	01.03.03	Isomerism of left atrial appendages	108224239	LA8Y	01.03.06	3	Isomerism of left atrial appendages	
13	13	02.04.12	Abnormal ventricular relationships	2036838536	LA81	03.01.13	4	Abnormal ventricular relationships	
14	14**	02.03.01	Right hand pattern ventricular topology	1541694179		02.14.12	1	Left-hand pattern ventricular topology	
15	15	02.03.02	Left hand pattern ventricular topology	1819755421	LA81	02.14.12	2	Right-hand pattern ventricular topology	
16	16	02.03.03	Crisscross heart	856695997	LA81	02.14.12	3	Crisscross heart (twisted atrioventricular connections)	Twisted atrioventricular connections
17	17	02.04.00	Superior-inferior ventricular relationship	1577251368	LA81	02.14.12	4	Superior-inferior ('upstairs-downstairs') ventricular relationship	'Upstairs-downstairs' ventricular relationship

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Table 3. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	ICD-11 Foundation entity number	ICD-11 MMS code or ICD-11 MMS crossmap	Parent	Sibling order	ICD-11 Congenital Cardiac term published in 2017[105] (terms in blue cells have been changed)	ICD-11 Congenital Cardiac term components published in 2017[105] and now moved to Synonyms
18	18	02.06.12	Abnormal relationship of great arterial roots	1403694832	LA8Y	03.01.13	5	Abnormal relationship of great arterial roots	
19	19	02.06.03	Aortic root directly anterior to pulmonary root	1491991321	LA8Y	02.06.12	1	Aortic root directly anterior to pulmonary root	
20	20	02.06.02	Aortic root anterior and rightward to pulmonary root	2054039932	LA8Y	02.06.12	2	Aortic root anterior and rightward to pulmonary root	
21	21	02.06.04	Aortic root anterior and leftward to pulmonary root	1308428290	LA8Y	02.06.12	3	Aortic root anterior and leftward to pulmonary root	
22	22	02.06.01	Aortic root side by side and directly rightward to pulmonary root	1682649237	LA8Y	02.06.12	4	Aortic root side by side and directly rightward to pulmonary root	
23	23	02.06.05	Aortic root side by side and directly leftward to pulmonary root	1447224468	LA8Y	02.06.12	5	Aortic root side by side and directly leftward to pulmonary root	
24	24	02.06.07	Aortic root directly posterior to pulmonary root	1772030433	LA8Y	02.06.12	6	Aortic root directly posterior to pulmonary root	
25	25**	02.06.00	Aortic root posterior and rightward to pulmonary root	1577855135		02.06.12	7	Aortic root posterior and rightward to pulmonary root	
26	26	02.06.06	Aortic root posterior and leftward to pulmonary root	1995160254	LA8Y	02.06.12	8	Aortic root posterior and leftward to pulmonary root	
27	27	02.07.03	Abnormal intrapericardial course of great arteries	1155690171	LA8Y	03.01.13	6	Abnormal intrapericardial course of great arteries	
28	28**	02.07.00	Spiralling course of great arteries	1834243628		02.07.03	1	Spiralling course of great arteries	
29	29	02.07.01	Parallel course of great arteries	1523021650	LA8Y	02.07.03	2	Parallel course of great arteries	
30	30	03.01.02	Visceral heterotaxy	780273165	LA8Y	03.01.13	7	Visceral heterotaxy (abnormal arrangement of thoraco-abdominal organs)	Abnormal arrangement of thoraco-abdominal organs
31	31	03.01.04	Right isomerism	1576694141	LA83	03.01.02	1	Right isomerism (‘asplenia syndrome’)	‘Asplenia syndrome’
32	32	03.01.05	Left Isomerism	1234712569	LA84	03.01.02	2	Left Isomerism (‘polysplenia syndrome’)	‘Polysplenia syndrome’

Table 3. (*Continued*)

33	33	03.01.03	Total mirror imagery	797648408	LA82	03.01.13	8	Total mirror imagery (situs inversus totalis)	Situs inversus totalis
34	34	01.03.09	Congenital anomaly of an atrioventricular or ventriculo-arterial connection	238916322	LA85	01.01.59	2	Congenital anomaly of an atrioventricular or ventriculo-arterial connection	
35	35**	01.04.00	Concordant atrioventricular connections	221514522		01.03.09	1	Concordant atrioventricular connections	
36	36	01.04.01	Discordant atrioventricular connections	230955773	LA85.0	01.03.09	2	Discordant atrioventricular connections	
37	37	01.01.03	Congenitally corrected transposition of great arteries	254915185	LA85.00	01.04.01	1	Congenitally corrected transposition of great arteries (discordant atrioventricular & ventriculo-arterial connections)	Discordant atrioventricular & ventriculo-arterial connections
38	38	01.05.01	Transposition of the great arteries	429190257	LA85.1	01.03.09	3	Transposition of the great arteries (discordant ventriculo-arterial connections)	Discordant ventriculo-arterial connections
39	39	01.01.02	Transposition of the great arteries with concordant atrioventricular connections and intact ventricular septum	1927702340	LA85.1	01.05.01	1	Transposition of the great arteries with concordant atrioventricular connections and intact ventricular septum	
40	40	01.01.10	Transposition of the great arteries with concordant atrioventricular connections and ventricular septal defect	1165313598	LA85.1	01.05.01	2	Transposition of the great arteries with concordant atrioventricular connections and ventricular septal defect	
41	41	01.01.10 + 07.09.01	Transposition of the great arteries with concordant atrioventricular connections and ventricular septal defect and left ventricular outflow tract obstruction	1744213081	LA85.1	01.05.01	3	Transposition of the great arteries with concordant atrioventricular connections and ventricular septal defect and left ventricular outflow tract obstruction	
42	42**	01.05.00	Concordant ventriculo-arterial connections	840265755		01.03.09	4	Concordant ventriculo-arterial connections	
43	43	01.05.10	Concordant ventriculo-arterial connections with parallel great arteries	1912928588	LA85.Y	01.05.00	1	Concordant ventriculo-arterial connections with parallel great arteries (anatomically corrected malposition)	Anatomically corrected malposition
44	44	01.01.04	Double outlet right ventricle	141717788	LA85.2	01.03.09	5	Double outlet right ventricle	
45	45	01.01.17	Double outlet right ventricle with subaortic or doubly committed	1060446859	LA85.23	01.01.04	1	Double outlet right ventricle with subaortic or doubly committed	

(*Continued*)

Table 3. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	ICD-11 Foundation entity number	ICD-11 MMS code or ICD-11 MMS crossmap	Parent	Sibling order	ICD-11 Congenital Cardiac term published in 2017[105] (terms in blue cells have been changed)	ICD-11 Congenital Cardiac term components published in 2017[105] and now moved to Synonyms
			ventricular septal defect and pulmonary stenosis, Fallot type					ventricular septal defect & pulmonary stenosis (Fallot type)	
46	46	01.01.17 + 07.13.04	Double outlet right ventricle with subaortic ventricular septal defect and pulmonary stenosis, Fallot type	2017764663	LA85.23	01.01.17	1	Double outlet right ventricle with subaortic ventricular septal defect & pulmonary stenosis	
47	47	01.01.17 + 07.13.02	Double outlet right ventricle with doubly committed ventricular septal defect and pulmonary stenosis, Fallot type	1460882984	LA85.23	01.01.17	2	Double outlet right ventricle with doubly committed ventricular septal defect & pulmonary stenosis	
48	48	01.01.18	Double outlet right ventricle with subpulmonary ventricular septal defect, transposition type	1963185163	LA85.20	01.01.04	2	Double outlet right ventricle with subpulmonary ventricular septal defect (transposition type)	
49	49	01.01.19	Double outlet right ventricle with non-committed ventricular septal defect	2032277111	LA85.21	01.01.04	3	Double outlet right ventricle with non-committed ventricular septal defect	
50	50	01.01.40	Double outlet right ventricle with subaortic or doubly committed ventricular septal defect without pulmonary stenosis, ventricular septal defect type	1410257155	LA85.22	01.01.04	4	Double outlet right ventricle with subaortic or doubly committed ventricular septal defect without pulmonary stenosis (ventricular septal defect type)	
51	51	01.01.40 + 07.13.04	Double outlet right ventricle with subaortic ventricular septal defect without pulmonary stenosis	434071545	LA85.22	01.01.40	1	Double outlet right ventricle with subaortic ventricular septal defect without pulmonary stenosis	
52	52	01.01.40 + 07.13.02	Double outlet right ventricle with doubly committed ventricular septal defect without pulmonary stenosis	776750727	LA85.22	01.01.40	2	Double outlet right ventricle with doubly committed ventricular septal defect without pulmonary stenosis	
53	53	01.01.24	Double outlet right ventricle with intact ventricular septum	1351035695	LA85.2Y	01.01.04	5	Double outlet right ventricle with intact ventricular septum	
54	54	01.05.03	Double outlet left ventricle	2094997989	LA85.3	01.03.09	6	Double outlet left ventricle	

Table 3. (*Continued*)

55	55	09.01.01	Common arterial trunk	1832500366	LA85.4	01.03.09	7	Common arterial trunk (Truncus arteriosus)	Truncus arteriosus
56	56	09.01.15	Common arterial trunk with aortic dominance	551770382	LA85.40	09.01.01	1	Common arterial trunk (truncus arteriosus) with aortic dominance (no aortic arch obstruction)	Truncus arteriosus with aortic dominance (no aortic arch obstruction)
57	57	09.01.14	Common arterial trunk with aortic dominance and both pulmonary arteries arising from trunk	214930658	LA85.40	09.01.15	1	Common arterial trunk (truncus arteriosus) with aortic dominance and both pulmonary arteries arising from trunk	Truncus arteriosus with aortic dominance and both pulmonary arteries arising from trunk
58	58	09.01.11	Common arterial trunk with aortic dominance and one pulmonary artery absent from the trunk, isolated pulmonary artery	524203135	LA85.40	09.01.15	2	Common arterial trunk (truncus arteriosus) with aortic dominance and one pulmonary artery absent from trunk (isolated pulmonary artery)	Truncus arteriosus with aortic dominance and one pulmonary artery absent from the trunk (isolated pulmonary artery)
59	59	09.01.12	Common arterial trunk with pulmonary dominance and aortic arch obstruction	659759166	LA85.4Y	09.01.01	2	Common arterial trunk (truncus arteriosus) with pulmonary dominance and aortic arch obstruction	Truncus arteriosus with pulmonary dominance and aortic arch obstruction
60	60	09.01.18	Common arterial trunk with pulmonary dominance and interrupted aortic arch	97579611	LA85.41	09.01.12	1	Common arterial trunk (truncus arteriosus) with pulmonary dominance and interrupted aortic arch	Truncus arteriosus with pulmonary dominance and interrupted aortic arch
61	61	09.01.19	Common arterial trunk with pulmonary dominance and aortic coarctation	843305765	LA85.4Y	09.01.12	2	Common arterial trunk (truncus arteriosus) with pulmonary dominance and aortic coarctation	Truncus arteriosus with pulmonary dominance and aortic coarctation
62	A^{+}	09.02.10	Atypical truncal valve	1983503623	LA85.4Y	09.01.01	3		
63	62	09.02.19	Congenital truncal valvar regurgitation	1421479778	LA85.4Y	09.02.10	1	Congenital truncal valvar regurgitation	
64	63	09.02.18	Congenital truncal valvar stenosis	2079188083	LA85.4Y	09.02.10	2	Congenital truncal valvar stenosis	
65	64	09.02.01	Dysplasia of truncal valve	1690328350	LA85.4Y	09.02.10	3	Dysplasia of truncal valve	
66	65	04.00.07	Congenital anomaly of mediastinal vein	1569064706	LA86	01.01.59	3	Congenital anomaly of mediastinal vein	
67	66	04.00.08	Congenital anomaly of mediastinal systemic vein	1524626968	LA86.Y	04.00.07	1	Congenital anomaly of mediastinal systemic vein	
68	67	04.01.09	Congenital anomaly of superior caval vein	1459500132	LA86.Y	04.00.08	1	Congenital anomaly of superior caval vein (superior vena cava)	Congenital anomaly of superior vena cava

(*Continued*)

Table 3. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	ICD-11 Foundation entity number	ICD-11 MMS code or ICD-11 MMS crossmap	Parent	Sibling order	ICD-11 Congenital Cardiac term published in 2017[105] (terms in blue cells have been changed)	ICD-11 Congenital Cardiac term components published in 2017[105] and now moved to Synonyms
69	68	04.01.05	Absent right superior caval vein	1665636689	LA86.Y	04.01.09	1	Absent right superior caval vein (superior vena cava)	Absent right superior vena cava
70	69	04.01.25	Left superior caval vein	804505819	LA86.0	04.01.09	2	Left superior caval vein (superior vena cava)	Left superior vena cava
71	70	04.01.01	Left superior caval vein to coronary sinus	523545228	LA86.0	04.01.25	1	Left superior caval vein (superior vena cava) to coronary sinus	Left superior vena cava to coronary sinus
72	71	04.01.02	Left superior caval vein to left-sided atrium	320170224	LA86.0	04.01.25	2	Left superior caval vein (superior vena cava) to left-sided atrium	Left superior vena cava to left-sided atrium
73	B^{+}	04.01.07	Congenital stenosis of superior caval vein	1640580264	LA86.Y	04.01.09	3		
74	72	04.03.08	Congenital anomaly of inferior caval vein	1266881625	LA86.Y	04.00.08	2	Congenital anomaly of inferior caval vein (inferior vena cava)	Congenital anomaly of inferior vena cava
75	73	04.03.10	Interrupted inferior caval vein with absent suprarenal segment and azygos continuation	1193646260	LA86.Y	04.03.08	1	Interrupted inferior caval vein (inferior vena cava) with absent suprarenal segment and azygos continuation	Interrupted inferior vena cava with absent suprarenal segment and azygos continuation
76	C^{+}	04.03.06	Congenital stenosis of inferior caval vein	100656181	LA86.Y	04.03.08	2		
77	74	04.04.05	Congenital anomaly of the coronary sinus	1737150764	LA86.Y	04.00.08	3	Congenital anomaly of coronary sinus	
78	75	04.04.13	Unroofed coronary sinus	800577917	LA86.1	04.04.05	1	Unroofed coronary sinus	
79	D^{+}	04.04.02	Completely unroofed coronary sinus	1900154411	LA86.1	04.04.13	1		
80	E^{+}	04.04.01	Partially unroofed coronary sinus	455093006	LA86.1	04.04.13	2		
81	76	04.04.14	Coronary sinus orifice atresia or stenosis	1460525276	LA86.Y	04.04.05	2	Coronary sinus orifice atresia or stenosis	
82	77***	04.02.13	Anomalous hepatic venous connection to heart	170873794	LB20.0Y	04.00.08	4	Anomalous hepatic venous connection to heart	
83	78	04.08.04	Congenital anomaly of pulmonary vein	969599265	LA86.Y	04.00.07	2	Congenital anomaly of pulmonary vein	
84	79	04.08.07	Anomalous pulmonary venous connection	1308345892	LA86.2	04.08.04	1	Anomalous pulmonary venous connection	

Table 3. (*Continued*)

85	80	04.08.05	Total anomalous pulmonary venous connection	1532925990	LA86.20	04.08.07	1	Total anomalous pulmonary venous connection
86	81	04.06.00	Total anomalous pulmonary venous connection of the supracardiac type	1914403600	LA86.20	04.08.05	1	Total anomalous pulmonary venous connection of supracardiac type
87	82	04.08.10	Total anomalous pulmonary venous connection of the cardiac type	2117777772	LA86.20	04.08.05	2	Total anomalous pulmonary venous connection of cardiac type
88	83	04.08.20	Total anomalous pulmonary venous connection of the infracardiac type	1784217576	LA86.20	04.08.05	3	Total anomalous pulmonary venous connection of infracardiac type
89	84	04.08.30	Total anomalous pulmonary venous connection of the mixed type	1719221652	LA86.20	04.08.05	4	Total anomalous pulmonary venous connection of mixed type
90	85	04.07.01	Partial anomalous pulmonary venous connection	1041585584	LA86.21	04.08.07	2	Partial anomalous pulmonary venous connection
91	86	01.01.16	Partial anomalous pulmonary venous connection of Scimitar type	211220931	LA86.2Y	04.08.07	3	Partial anomalous pulmonary venous connection of Scimitar type
92	87	03.02.23	Scimitar syndrome	1321054364	LA86.22	01.01.16	1	Scimitar syndrome
93	88	04.08.06	Obstructed anomalous pulmonary venous pathway or connection	1577171884	LA86.2Y	04.08.07	4	Obstructed anomalous pulmonary venous pathway or connection
94	89	04.08.31	Congenital pulmonary venous stenosis or hypoplasia	469101490	LA86.3	04.08.04	2	Congenital pulmonary venous stenosis and/or hypoplasia
95	90	04.08.02	Congenital atresia of pulmonary vein	1937562872	LA86.Y	04.08.04	3	Congenital atresia of pulmonary vein
96	91	05.00.02	Congenital anomaly of an atrium or atrial septum	92905340	LA8Y	01.01.59	4	Congenital anomaly of an atrium and/or atrial septum
97	92	05.07.01	Congenital anomaly of atrial septum	654986527	LA8E	05.00.02	1	Congenital anomaly of atrial septum
98	93	05.06.04	Restrictive interatrial communication or intact atrial septum when an interatrial shunt is physiologically necessary	499082002	LA8E.Y	05.07.01	1	Restrictive interatrial communication or intact atrial septum when an interatrial shunt is physiologically necessary
99	94	05.03.03	Aneurysm of atrial septum	100700036	LA8E.Y	05.07.01	2	Aneurysm of atrial septum

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Table 3. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	ICD-11 Foundation entity number	ICD-11 MMS code or ICD-11 MMS crossmap	Parent	Sibling order	ICD-11 Congenital Cardiac term published in 2017[105] (terms in blue cells have been changed)	ICD-11 Congenital Cardiac term components published in 2017[105] and now moved to Synonyms
100	95	05.04.01	Interatrial communication	1285985084	LA8E.Y	05.07.01	3	Interatrial communication ('atrial septal defect')	'Atrial septal defect'
101	96**	05.03.01	Patent oval foramen	1618980674	LA8E.0	05.04.01	1	Patent oval foramen (patent foramen ovale)	Patent foramen ovale
102	97	05.04.02	Atrial septal defect within oval fossa	1875768490	LA8E.1	05.04.01	2	Atrial septal defect within oval fossa (secundum atrial septal defect)	Secundum atrial septal defect
103	98	05.05.00	Sinus venosus defect	1930019148	LA8E.2	05.04.01	3	Sinus venosus defect	
104	99	05.06.02	Common atrium with separate atrioventricular junctions	1022267780	LA8E.Y	05.04.01	4	Common atrium with separate atrioventricular junctions	
105	100	05.05.03	Interatrial communication through coronary sinus orifice	664625334	LA8E.3	05.04.01	5	Interatrial communication through coronary sinus orifice	
106	101	05.01.13	Congenital anomaly of right atrium	1523246177	LA8F	05.00.02	2	Congenital anomaly of right atrium	
107	102	05.01.21	Divided right atrium	294192583	LA8F	05.01.13	1	Divided right atrium (cor triatriatum dexter)	Cor triatriatum dexter
108	F+	05.01.04	Chiari network	898914868	LA8F	05.01.13	2		
109	103	05.01.06	Left-sided juxtaposition of the atrial appendages	1461640469	LA8F	05.01.13	3	Left-sided juxtaposition of atrial appendages	
110	104	05.01.12	Congenital giant right atrium	2067836926	LA8F	05.01.13	4	Congenital giant right atrium	
111	105	05.02.11	Congenital anomaly of left atrium	1757622374	LA8G	05.00.02	3	Congenital anomaly of left atrium	
112	106	05.02.01	Divided left atrium	90967508	LA8G.0	05.02.11	1	Divided left atrium (cor triatriatum sinister)	Cor triatriatum sinister
113	107	05.02.04	Right-sided juxtaposition of the atrial appendages	1619917931	LA8G.Y	05.02.11	2	Right-sided juxtaposition of atrial appendages	
114	108	06.00.15	Congenital anomaly of an atrioventricular valve or atrioventricular septum	1055878726	LA87	01.01.59	5	Congenital anomaly of an atrioventricular valve and/or atrioventricular septum	
115	109	06.01.11	Congenital anomaly of tricuspid valve	995525654	LA87.0	06.00.15	1	Congenital anomaly of tricuspid valve	
116	110	06.01.25	Congenital tricuspid regurgitation	1523583011	LA87.00	06.01.11	1	Congenital tricuspid regurgitation	
117	111	06.01.07	Congenital tricuspid valvar stenosis	1996822362	LA87.01	06.01.11	2	Congenital tricuspid valvar stenosis	

Table 3. (*Continued*)

118	112	06.01.04	Tricuspid annular hypoplasia	281173100	LA87.0Y	06.01.11	3	Tricuspid annular hypoplasia	
119	113	06.01.03	Dysplasia of tricuspid valve	1468235714	LA87.02	06.01.11	4	Dysplasia of tricuspid valve	
120	114	06.01.09	Straddling tricuspid valve	908739636	LA87.0Y	06.01.11	5	Straddling tricuspid valve	
121	115	06.01.05	Overriding tricuspid valve	1584596137	LA87.0Y	06.01.11	6	Overriding tricuspid valve	
122	116	06.01.34	Ebstein malformation of tricuspid valve	307157712	LA87.03	06.01.11	7	Ebstein malformation of tricuspid valve	
123	G^{+}	06.01.32	Absent tricuspid valve leaflet	603292687	LA87.0Y	06.01.11	8		
124	H^{+}	06.01.36	True cleft of tricuspid valve leaflet	1733245532	LA87.0Y	06.01.11	9		
125	117	06.02.11	Congenital anomaly of mitral valve	928274753	LA87.1	06.00.15	2	Congenital anomaly of mitral valve	
126	118	06.02.25	Congenital mitral regurgitation	403917903	LA87.10	06.02.11	1	Congenital mitral regurgitation	
127	119	06.02.07	Congenital mitral valvar stenosis	2102952411	LA87.11	06.02.11	2	Congenital mitral valvar stenosis	
128	120	06.02.04	Mitral annular hypoplasia	843702959	LA87.1Y	06.02.11	3	Mitral annular hypoplasia	
129	121	06.02.09	Straddling mitral valve	930563690	LA87.1Y	06.02.11	4	Straddling mitral valve	
130	122	06.02.05	Overriding mitral valve	462079876	LA87.1Y	06.02.11	5	Overriding mitral valve	
131	123	06.02.03	Dysplasia of mitral valve	1958615745	LA87.12	06.02.11	6	Dysplasia of mitral valve	
132	124	05.02.02	Supravalvar or intravalvar mitral ring	1868985430	LA87.1Y	06.02.11	7	Supravalvar or intravalvar mitral ring	
133	I^{+}	06.02.23	Congenital intravalvar mitral ring	594666245	LA87.1Y	05.02.02	1		
134	J^{+}	06.02.17	Congenital supravalvar mitral ring	308712619	LA87.1Y	05.02.02	2		
135	125	06.02.72	Congenital mitral valvar prolapse	840405955	LA87.1Y	06.02.11	8	Congenital mitral valvar prolapse	
136	126	06.02.36	True cleft of anterior mitral leaflet	250037637	LA87.1Y	06.02.11	9	True cleft of anterior mitral leaflet (without common atrioventricular junction)	True cleft of anterior mitral leaflet (without common atrioventricular junction)
137	127	06.02.21	Congenital anomaly of mitral subvalvar apparatus	498751490	LA87.13	06.02.11	10	Congenital anomaly of mitral subvalvar apparatus	
138	128	06.02.22	Congenital mitral subvalvar stenosis	659651858	LA87.13	06.02.21	1	Congenital mitral subvalvar stenosis	
139	129	06.02.56	Parachute malformation of mitral valve	375324402	LA87.13	06.02.21	2	Parachute malformation of mitral valve	

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Table 3. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	ICD-11 Foundation entity number	ICD-11 MMS code or ICD-11 MMS crossmap	Parent	Sibling order	ICD-11 Congenital Cardiac term published in 2017[105] (terms in blue cells have been changed)	ICD-11 Congenital Cardiac term components published in 2017[105] and now moved to Synonyms
140	K+	06.02.39	Accessory tissue on mitral valve leaflet	248264200	LA87.1Y	06.02.11	11		
141	L+	06.02.32	Congenital unguarded mitral orifice	1824064279	LA87.1Y	06.02.11	12		
142	M+	06.02.33	Double orifice of mitral valve	1045118968	LA87.1Y	06.02.11	13		
143	130	06.04.11	Congenital anomaly of left-sided atrioventricular valve in double inlet ventricle	602975543	LA87.Y	06.00.15	5	Congenital anomaly of left-sided atrioventricular valve in double inlet ventricle	
144	131	06.03.11	Congenital anomaly of right-sided atrioventricular valve in double inlet ventricle	122794405	LA87.Y	06.00.15	4	Congenital anomaly of right-sided atrioventricular valve in double inlet ventricle	
145	132	06.06.11	Common atrioventricular junction	1729725342	LA87.Y	06.00.15	3	Common atrioventricular junction (common atrioventricular canal)	Common atrioventricular canal
146	133	06.06.00	Common atrioventricular junction with atrioventricular septal defect	1613228388	LA87.2	06.06.11	1	Atrioventricular septal defect (atrioventricular canal defect)	Atrioventricular canal defect
147	134	06.07.27	Atrioventricular septal defect with balanced ventricles	831762174	LA87.2Y	06.06.00	1	Atrioventricular septal defect with balanced ventricles	
148	135	06.07.26	Atrioventricular septal defect with ventricular imbalance	560731030	LA87.25	06.06.00	2	Atrioventricular septal defect with ventricular imbalance	
149	136	06.07.05	Atrioventricular septal defect with ventricular imbalance with dominant right ventricle and hypoplastic left ventricle	66044403	LA87.25	06.07.26	1	Atrioventricular septal defect with ventricular imbalance: dominant right ventricle, hypoplastic left ventricle	
150	137	06.07.06	Atrioventricular septal defect with ventricular imbalance with dominant left ventricle and hypoplastic right ventricle	880599181	LA87.25	06.07.26	2	Atrioventricular septal defect with ventricular imbalance: dominant left ventricle, hypoplastic right ventricle	
151	138	06.06.01	Atrioventricular septal defect with communication at the atrial level only	1159570489	LA87.21	06.06.00	3	Atrioventricular septal defect with communication at the atrial level only (primum atrial septal defect) (partial atrioventricular canal defect)	Primum atrial septal defect (Partial atrioventricular canal defect)

Table 3. (*Continued*)

152	139	06.06.08	Atrioventricular septal defect with communication at the ventricular level only	793233560	LA87.22	06.06.00	4	Atrioventricular septal defect with communication at the ventricular level only (atrioventricular canal defect with isolated ventricular communication)	Atrioventricular canal defect with isolated ventricular communication
153	140	06.06.10	Atrioventricular septal defect with communication at atrial level and restrictive communication at ventricular level	687321516	LA87.23	06.06.00	5	Atrioventricular septal defect (atrioventricular canal defect) with communication at atrial level and restrictive communication at ventricular level (intermediate atrioventricular septal defect) (transitional atrioventricular septal defect)	Intermediate atrioventricular septal defect (Transitional atrioventricular septal defect)
154	141	06.06.09	Atrioventricular septal defect with communication at atrial level and unrestrictive communication at ventricular level	551617570	LA87.24	06.06.00	6	Atrioventricular septal defect (atrioventricular canal defect) with communication at atrial level and unrestrictive communication at ventricular level (Complete atrioventricular septal defect) (Complete atrioventricular canal defect)	Complete atrioventricular septal defect (Complete atrioventricular canal defect)
155	142	01.01.20	Atrioventricular septal defect and tetralogy of Fallot	915281294	LA87.2Y	06.06.00	7	Atrioventricular septal defect and tetralogy of Fallot (atrioventricular canal and tetralogy of Fallot)	Atrioventricular canal and tetralogy of Fallot
156	143	05.06.03	Common atrium with common atrioventricular junction	240010127	LA87.2Y	06.06.00	8	Common atrium with common atrioventricular junction	
157	144	06.05.60	Common atrioventricular valvar regurgitation	908318670	LA87.2Y	06.06.00	9	Common atrioventricular valvar regurgitation	
158	145	06.05.14	Atypical common atrioventricular valve	346031531	LA87.2Y	06.06.00	10	Atypical common atrioventricular valve	
159	146	06.07.36	Common atrioventricular valve with unbalanced commitment of valve to ventricles	590554325	LA87.2Y	06.05.14	1	Common atrioventricular valve with unbalanced commitment of valve to ventricles	
160	147	06.07.37	Common atrioventricular valve with unbalanced commitment of valve to right ventricle	176495682	LA87.2Y	06.07.36	1	Common atrioventricular valve with unbalanced commitment of valve to right ventricle	
161	148	06.07.38	Common atrioventricular valve with unbalanced commitment of valve to left ventricle	948750405	LA87.2Y	06.07.36	2	Common atrioventricular valve with unbalanced commitment of valve to left ventricle	
162	149	06.05.71	Atypical right ventricular component of common atrioventricular valve	122758456	LA87.2Y	06.05.14	2	Atypical right ventricular component of common atrioventricular valve	

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Table 3. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	ICD-11 Foundation entity number	ICD-11 MMS code or ICD-11 MMS crossmap	Parent	Sibling order	ICD-11 Congenital Cardiac term published in 2017[105] (terms in blue cells have been changed)	ICD-11 Congenital Cardiac term components published in 2017[105] and now moved to Synonyms
163	150	06.05.72	Atypical left ventricular component of common atrioventricular valve	818021450	LA87.2Y	06.05.14	3	Atypical left ventricular component of common atrioventricular valve	
164	151	06.05.25	Double orifice of left ventricular component of common atrioventricular valve	1297798321	LA87.2Y	06.05.72	1	Double orifice of left ventricular component of common atrioventricular valve	
165	152	06.05.98	Deficient mural leaflet of left ventricular component of common atrioventricular valve	2136146102	LA87.2Y	06.05.72	2	Deficient mural (lateral) leaflet of left ventricular component of common atrioventricular valve	Deficient lateral leaflet of left ventricular component of common atrioventricular valve
166	153	06.07.28	Common atrioventricular junction without an atrioventricular septal defect	1457862689	LA87.3	06.06.11	2	Common atrioventricular junction with spontaneous fibrous closure of atrioventricular septal defect	
167	154	07.14.02	Communication between left ventricle and right atrium	1370033158	LA87.Y	06.00.15	6	Communication between left ventricle and right atrium (Gerbode defect)	Gerbode defect
168	155	07.00.00	Congenital anomaly of a ventricle or the ventricular septum	508003685	LA88	01.01.59	6	Congenital anomaly of a ventricle and/or the ventricular septum	
169	156	07.01.07	Congenital right ventricular anomaly	1019500288	LA88.Y	07.00.00	1	Congenital right ventricular anomaly	
170	157	07.02.00	Right ventricular hypoplasia	2088837187	LA88.Y	07.01.07	1	Right ventricular hypoplasia	
171	158	07.05.20	Congenital right ventricular outflow tract obstruction	1259471165	LA88.0	07.01.07	2	Congenital right ventricular outflow tract obstruction	
172	159	07.03.01	Double chambered right ventricle	997469748	LA88.1	07.01.07	3	Double chambered right ventricle	
173	160	07.01.13	Right ventricular myocardial sinusoids	1293423457	LA88.Y	07.01.07	4	Right ventricular myocardial sinusoids	
174	161	07.01.06	Parchment right ventricle	240652322	LA88.Y	07.01.07	5	Parchment right ventricle (including Uhl anomaly)	Parchment right ventricle (including Uhl anomaly)
175	162	01.01.01	Tetralogy of Fallot	90973426	LA88.2	07.01.07	6	Tetralogy of Fallot	
176	163	09.05.25	Tetralogy of Fallot with absent pulmonary valve syndrome	1640350515	LA88.20	01.01.01	1	Tetralogy of Fallot with absent pulmonary valve syndrome	

Table 3. (*Continued*)

177	164	01.01.26	Tetralogy of Fallot with pulmonary atresia	1645917296	LA88.21	01.01.01	2	Tetralogy of Fallot with pulmonary atresia	
178	165	01.01.57	Tetralogy of Fallot with pulmonary atresia and systemic-to-pulmonary collateral arteries	1408174111	LA88.22	01.01.01	3	Tetralogy of Fallot with pulmonary atresia and systemic-to-pulmonary collateral artery(ies)	Tetralogy of Fallot with pulmonary atresia and systemic-to-pulmonary collateral artery(ies)
179	166	07.06.07	Congenital left ventricular anomaly	328449041	L88.Y	07.00.00	2	Congenital left ventricular anomaly	
180	167	07.07.00	Left ventricular hypoplasia	1353575853	L88.Y	07.06.07	1	Left ventricular hypoplasia	
181	168	07.06.19	Congenital left ventricular aneurysm or diverticulum	208890733	L88.Y	07.06.07	2	Congenital left ventricular aneurysm or diverticulum	
182	N+	07.06.01	Congenital left ventricular aneurysm	416161260	L88.Y	07.06.19	1		
183	O+	07.06.03	Congenital left ventricular diverticulum	378873520	L88.Y	07.06.19	2		
184	169	07.09.28	Congenital left ventricular outflow tract obstruction	567908339	LA88.3	07.06.07	3	Congenital left ventricular outflow tract obstruction	
185	170	07.09.08	Congenital left ventricular outflow tract obstruction due to atrioventricular valve	1869611230	LA88.3	07.09.28	1	Left ventricular outflow tract obstruction due to atrioventricular valve	
186	171	01.01.33	Left heart obstruction at multiple sites	295410302	LA88.Y	07.06.07	4	Left heart obstruction at multiple sites (including Shone syndrome)	Left heart obstruction at multiple sites (including Shone syndrome)
187	172	07.06.12	Left ventricular myocardial sinusoids	1635129695	LA88.Y	07.06.07	5	Left ventricular myocardial sinusoids	
188	P+	07.00.07	Anomalous ventricular bands	1260829039	LA88.Y	07.00.00	4		
189	173	07.20.04	Congenital anomaly of ventricular septum	1908503567	LA88.Y	07.00.00	3	Congenital anomaly of ventricular septum	
190	174	07.14.07	Restrictive interventricular communication when an interventricular shunt is physiologically necessary	1822564395	LA88.Y	07.20.04	1	Restrictive interventricular communication when an interventricular shunt is physiologically necessary	
191	175	07.10.00	Ventricular septal defect	668140715	LA88.4	07.20.04	2	Ventricular septal defect	
192	176	07.10.01	Perimembranous central ventricular septal defect	2023258628	LA88.41	07.10.00	1	Perimembranous central ventricular septal defect	
193	177	07.14.05	Inlet ventricular septal defect without a common atrioventricular junction	1491280288	LA88.4Y	07.10.00	2	Inlet ventricular septal defect without a common atrioventricular junction	

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Table 3. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	ICD-11 Foundation entity number	ICD-11 MMS code or ICD-11 MMS crossmap	Parent	Sibling order	ICD-11 Congenital Cardiac term published in 2017[105] (terms in blue cells have been changed)	ICD-11 Congenital Cardiac term components published in 2017[105] and now moved to Synonyms
194	178	07.10.02	Inlet perimembranous ventricular septal defect without atrioventricular septal malalignment without a common atrioventricular junction	502334750	LA88.4Y	07.14.05	1	Inlet perimembranous ventricular septal defect without atrioventricular septal malalignment without a common atrioventricular junction	
195	179	07.14.06	Inlet perimembranous ventricular septal defect with atrioventricular septal malalignment and without a common atrioventricular junction	801142160	LA88.4Y	07.14.05	2	Inlet perimembranous ventricular septal defect with atrioventricular septal malalignment and without a common atrioventricular junction	
196	180	07.11.02	Inlet muscular ventricular septal defect	1145418250	LA88.4Y	07.14.05	3	Inlet muscular ventricular septal defect	
197	181	07.11.01	Trabecular muscular ventricular septal defect	65805952	LA88.40	07.10.00	3	Trabecular muscular ventricular septal defect	
198	182	07.11.04	Trabecular muscular ventricular septal defect midseptal	1131685234	LA88.40	07.11.01	1	Trabecular muscular ventricular septal defect: midseptal	
199	183	07.11.03	Trabecular muscular ventricular septal defect apical	1256893827	LA88.40	07.11.01	2	Trabecular muscular ventricular septal defect: apical	
200	184	07.11.12	Trabecular muscular ventricular septal defect postero-inferior	601485273	LA88.40	07.11.01	3	Trabecular muscular ventricular septal defect: postero-inferior	
201	185	07.11.07	Trabecular muscular ventricular septal defect anterosuperior	1220890087	LA88.40	07.11.01	4	Trabecular muscular ventricular septal defect: anterosuperior	
202	186	07.11.05	Multiple trabecular muscular ventricular septal defects	2105117949	LA88.40	07.11.01	5	Trabecular muscular ventricular septal defect: multiple (“Swiss cheese” septum)	“Swiss cheese” septum
203	187	07.12.00	Outlet ventricular septal defect	1879356291	LA88.4Y	07.10.00	4	Outlet ventricular septal defect	
204	188	07.12.09	Outlet ventricular septal defect without malalignment	7379274	LA88.4Y	07.12.00	1	Outlet ventricular septal defect without malalignment	
205	189	07.11.06	Outlet muscular ventricular septal defect without malalignment	222010536	LA88.4Y	07.12.09	1	Outlet muscular ventricular septal defect without malalignment	

Table 3. (*Continued*)

206	190	07.12.01	Doubly committed juxta-arterial ventricular septal defect without malalignment	1767950841	LA88.4Y	07.12.09	2	Doubly committed juxta-arterial ventricular septal defect without malalignment
207	191	07.12.02	Doubly committed juxta-arterial ventricular septal defect without malalignment and with muscular postero-inferior rim	1281801530	LA88.4Y	07.12.01	1	Doubly committed juxta-arterial ventricular septal defect without malalignment and with muscular postero-inferior rim
208	192	07.12.03	Doubly committed juxta-arterial ventricular septal defect without malalignment and with perimembranous extension	1519452698	LA88.4Y	07.12.01	2	Doubly committed juxta-arterial ventricular septal defect without malalignment and with perimembranous extension
209	193	07.10.17	Outlet ventricular septal defect with anteriorly malaligned outlet septum	1377580315	LA88.4Y	07.12.00	2	Outlet ventricular septal defect with anteriorly malaligned outlet septum
210	194	07.11.15	Outlet muscular ventricular septal defect with anteriorly malaligned outlet septum	391701468	LA88.4Y	07.10.17	1	Outlet muscular ventricular septal defect with anteriorly malaligned outlet septum
211	195	07.10.04	Outlet perimembranous ventricular septal defect with anteriorly malaligned outlet septum	1560415737	LA88.4Y	07.10.17	2	Outlet perimembranous ventricular septal defect with anteriorly malaligned outlet septum
212	196	07.12.12	Doubly committed juxta-arterial ventricular septal defect with anteriorly malaligned fibrous outlet septum	1226705023	LA88.4Y	07.10.17	3	Doubly committed juxta-arterial ventricular septal defect with anteriorly malaligned fibrous outlet septum
213	197	07.12.07	Doubly committed juxta-arterial ventricular septal defect with anteriorly malaligned fibrous outlet septum and muscular postero-inferior rim	1193651839	LA88.4Y	07.12.12	1	Doubly committed juxta-arterial ventricular septal defect with anteriorly malaligned fibrous outlet septum and muscular postero-inferior rim
214	198	07.12.05	Doubly committed juxta-arterial ventricular septal defect with anteriorly malaligned fibrous outlet septum and perimembranous extension	1496410457	LA88.4Y	07.12.12	2	Doubly committed juxta-arterial ventricular septal defect with anteriorly malaligned fibrous outlet septum and perimembranous extension
215	199	07.10.18	Outlet ventricular septal defect with posteriorly malaligned outlet septum	1478451433	LA88.4Y	07.12.00	3	Outlet ventricular septal defect with posteriorly malaligned outlet septum

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Table 3. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	ICD-11 Foundation entity number	ICD-11 MMS code or ICD-11 MMS crossmap	Parent	Sibling order	ICD-11 Congenital Cardiac term published in 2017[105] (terms in blue cells have been changed)	ICD-11 Congenital Cardiac term components published in 2017[105] and now moved to Synonyms
216	200	07.11.16	Outlet muscular ventricular septal defect with posteriorly malaligned outlet septum	903557561	LA88.4Y	07.10.18	1	Outlet muscular ventricular septal defect with posteriorly malaligned outlet septum	
217	201	07.10.19	Outlet perimembranous ventricular septal defect with posteriorly malaligned outlet septum	399222458	LA88.4Y	07.10.18	2	Outlet perimembranous ventricular septal defect with posteriorly malaligned outlet septum	
218	202	07.12.13	Doubly committed juxta-arterial ventricular septal defect with posteriorly malaligned fibrous outlet septum	1654184537	LA88.4Y	07.10.18	3	Doubly committed juxta-arterial ventricular septal defect with posteriorly malaligned fibrous outlet septum	
219	203	07.12.08	Doubly committed juxta-arterial ventricular septal defect with posteriorly malaligned fibrous outlet septum and muscular postero-inferior rim	802528118	LA88.4Y	07.12.13	1	Doubly committed juxta-arterial ventricular septal defect with posteriorly malaligned fibrous outlet septum and muscular postero-inferior rim	
220	204	07.12.06	Doubly committed juxta-arterial ventricular septal defect with posteriorly malaligned fibrous outlet septum and perimembranous extension	1207056363	LA88.4Y	07.12.13	2	Doubly committed juxta-arterial ventricular septal defect with posteriorly malaligned fibrous outlet septum and perimembranous extension	
221	205	07.15.01	Ventricular septal defect haemodynamically insignificant	924554858	LA88.42	07.10.00	5	Ventricular septal defect: Hemodynamically insignificant	
222	206	07.15.04	Multiple ventricular septal defects	1569246842	LA88.4Y	07.10.00	6	Multiple ventricular septal defects	
223	207	01.01.22	Functionally univentricular heart	5417233	LA89	01.01.59	7	Functionally univentricular heart	
224	208	01.01.14	Double inlet atrioventricular connection	1786413029	LA89.0	01.01.22	1	Double inlet atrioventricular connection (double inlet ventricle)	Double inlet ventricle
225	209	01.04.04	Double inlet left ventricle	568907539	LA89.0	01.01.14	1	Double inlet left ventricle	
226	210	01.04.03	Double inlet right ventricle	333513492	LA89.0	01.01.14	2	Double inlet right ventricle	
227	211	01.04.05	Double inlet to solitary ventricle of indeterminate morphology	1239626873	LA89.0	01.01.14	3	Double inlet to solitary ventricle of indeterminate morphology	
228	212	06.01.01	Tricuspid atresia	845891723	LA89.1	01.01.22	2	Tricuspid atresia	

Table 3. (*Continued*)

229	213	06.01.26	Tricuspid atresia with absent atrioventricular connection	60494995	LA89.1	06.01.01	1	Tricuspid atresia with absent valvar annulus (connection/junction)	Tricuspid atresia with absent valvar annulus (connection/junction)
230	214	06.01.02	Tricuspid atresia with imperforate tricuspid valve	1011284448	LA89.1	06.01.01	2	Tricuspid atresia with imperforate tricuspid valve	
231	215	06.02.01	Mitral atresia	6462604	LA89.2	01.01.22	3	Mitral atresia	
232	216	06.02.26	Mitral atresia with absent atrioventricular connection	198563998	LA89.2	06.02.01	1	Mitral atresia with absent valvar annulus (connection/junction)	Mitral atresia with absent valvar annulus (connection/junction)
233	217	06.02.02	Mitral atresia with imperforate mitral valve	590498493	LA89.2	06.02.01	2	Mitral atresia with imperforate mitral valve	
234	218	01.01.09	Hypoplastic left heart syndrome	1811800027	LA89.3	01.01.22	4	Hypoplastic left heart syndrome	
235	219	09.04.29	Congenital anomaly of a ventriculo-arterial valve or adjacent regions	1691908317	LA8A	01.01.59	8	Congenital anomaly of a ventriculo-arterial valve and/or adjacent regions	
236	220	09.05.29	Congenital anomaly of pulmonary valve	1754792043	LA8A.0	09.04.29	1	Congenital anomaly of pulmonary valve	
237	221	09.05.04	Congenital pulmonary valvar stenosis	353180069	LA8A.00	09.05.29	1	Congenital pulmonary valvar stenosis	
238	222	09.05.05	Pulmonary annular hypoplasia	663601170	LA8A.0Y	09.05.29	2	Pulmonary ‘annular’ hypoplasia	Pulmonary ‘annular’ hypoplasia
239	223	09.05.22	Congenital pulmonary regurgitation	1637894492	LA8A.01	09.05.29	3	Congenital pulmonary regurgitation	
240	224	09.05.24	Dysplasia of pulmonary valve	706270871	LA8A.0Y	09.05.29	4	Dysplasia of pulmonary valve	
241	225	09.05.32	Bicuspid pulmonary valve	1243337232	LA8A.0Y	09.05.29	5	Bicuspid pulmonary valve	
242	226	07.05.32	Congenital subpulmonary stenosis	1393194578	LA8A.6	09.04.29	2	Congenital subpulmonary stenosis	
243	227	09.07.15	Congenital supravalvar pulmonary stenosis	1801512478	LA8A.Y	09.04.29	3	Congenital supravalvar pulmonary stenosis	
244	228	09.05.16	Congenital pulmonary atresia	1883690033	LA8A.1	09.04.29	4	Congenital pulmonary atresia	
245	Q+	09.05.12	Congenital pulmonary valvar atresia	2133636301	LA8A.1Y	09.05.16	1		
246	229	01.01.07	Pulmonary atresia with intact ventricular septum	131289265	LA8A.10	09.05.16	2	Pulmonary atresia with intact ventricular septum	
247	230	09.15.19	Congenital anomaly of aortic valve	1932996411	LA8A.2	09.04.29	5	Congenital anomaly of aortic valve	

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Table 3. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	ICD-11 Foundation entity number	ICD-11 MMS code or ICD-11 MMS crossmap	Parent	Sibling order	ICD-11 Congenital Cardiac term published in 2017[105] (terms in blue cells have been changed)	ICD-11 Congenital Cardiac term components published in 2017[105] and now moved to Synonyms
248	231	09.15.01	Congenital aortic valvar stenosis	1824398514	LA8A.20	09.15.19	1	Congenital aortic valvar stenosis	
249	232	09.15.07	Congenital aortic regurgitation	167104804	LA8A.21	09.15.19	2	Congenital aortic regurgitation	
250	233	09.15.22	Bicuspid aortic valve	1328968452	LA8A.22	09.15.19	3	Bicuspid aortic valve	
251	234	09.15.21	Unicuspid aortic valve	725458981	LA8A.24	09.15.19	4	Unicuspid aortic valve	
252	235***	09.15.30	Aortic valvar prolapse	4999832858	BB74	09.15.19	5	Aortic valvar prolapse	
253	236	09.15.06	Aortic valvar atresia	1700740306	LA8A.23	09.15.19	6	Aortic valvar atresia	
254	237	09.15.17	Aortic annular hypoplasia	801311773	LA8A.2Y	09.15.19	7	Aortic ‘annular’ hypoplasia	Aortic ‘annular’ hypoplasia
255	238	09.15.09	Dysplasia of aortic valve	185322419	LA8A.2Y	09.15.19	8	Dysplasia of aortic valve	
256	239	07.09.50	Congenital subaortic stenosis	1350872731	LA8A.5	09.04.29	6	Congenital subaortic stenosis	
257	240	07.09.03	Subaortic stenosis due to fibromuscular shelf	1420460223	LA8A.5	07.09.50	1	Subaortic stenosis due to fibromuscular shelf	
258	241	07.09.16	Subaortic stenosis due to fibromuscular tunnel	159716932	LA8A.5	07.09.50	2	Subaortic stenosis due to fibromuscular tunnel	
259	242	09.16.18	Congenital supravalvar aortic stenosis	1066595728	LA8A.3	09.04.29	9	Congenital supravalvar aortic stenosis	
260	243	09.18.01	Aneurysm of aortic sinus of Valsalva	364348641	LA8A.4	09.04.29	10	Aneurysm of aortic sinus of Valsalva	
261	244	09.17.01	Aortoventricular tunnel	470594532	LA8A.Y	09.04.29	11	Aortoventricular tunnel	
262	R^{+}	09.17.02	Aorto-left ventricular tunnel	1781288740	LA8A.Y	09.17.01	1		
263	S^{+}	09.17.04	Aorto-right ventricular tunnel	1166626076	LA8A.Y	09.17.01	2		
264	245	09.04.28	Congenital anomaly of great arteries including arterial duct	1851979900	LA8B	01.01.59	9	Congenital anomaly of great arteries including arterial duct	
265	246	09.04.07	Congenital aortopulmonary window	1988278118	LA8B.0	09.04.28	1	Congenital aortopulmonary window	
266	247	09.07.16	Congenital anomaly of pulmonary arterial tree	953235173	LA8B.1	09.04.28	2	Congenital anomaly of pulmonary arterial tree	
267	248	09.10.36	Congenital dilation of pulmonary arterial tree	2022819457	LA8B.1	09.07.16	1	Congenital dilation of pulmonary arterial tree	

Table 3. (*Continued*)

268	249	09.07.19	Congenital pulmonary trunk anomaly	1325590527	LA8B.1	09.07.16	2	Congenital pulmonary trunk (main pulmonary artery) anomaly	Congenital main pulmonary artery anomaly
269	250	09.07.20	Congenital pulmonary trunk hypoplasia	1152699781	LA8B.1	09.07.19	1	Congenital pulmonary trunk (main pulmonary artery) hypoplasia	Congenital main pulmonary artery hypoplasia
270	251	09.07.05	Absent or atretic pulmonary trunk	1075480254	LA8B.1	09.07.19	2	Absent or atretic pulmonary trunk (main pulmonary artery)	Absent or atretic main pulmonary artery
271	252	09.10.41	Congenital pulmonary arterial branch anomaly	1743091164	LA8B.1	09.07.16	3	Congenital pulmonary arterial (branch) anomaly	
272	253	09.10.27	Congenital pulmonary arterial branch stenosis	1441571291	LA8B.1	09.10.41	1	Congenital pulmonary arterial (branch) stenosis	
273	254	09.10.28	Congenital right pulmonary arterial stenosis	2086312270	LA8B.1	09.10.27	1	Congenital right pulmonary arterial (branch) stenosis	Congenital right pulmonary arterial (branch) stenosis
274	255	09.10.29	Congenital left pulmonary arterial stenosis	1271511092	LA8B.1	09.10.27	2	Congenital left pulmonary arterial (branch) stenosis	Congenital left pulmonary arterial (branch) stenosis
275	256	09.10.71	Congenital pulmonary arterial branch hypoplasia	1766484567	LA8B.1	09.10.41	2	Congenital pulmonary arterial (branch) hypoplasia	
276	257	09.10.72	Congenital right pulmonary arterial hypoplasia	1176338758	LA8B.1	09.10.71	1	Congenital right pulmonary arterial (branch) hypoplasia	Congenital right pulmonary arterial (branch) hypoplasia
277	258	09.10.73	Congenital left pulmonary arterial hypoplasia	147957913	LA8B.1	09.10.71	2	Congenital left pulmonary arterial (branch) hypoplasia	Congenital left pulmonary arterial (branch) hypoplasia
278	T^{+}	09.10.21	Absent or atretic right or left pulmonary artery	542905766	LA8B.1	09.10.41	3		
279	259	09.10.75	Absent or atretic right pulmonary artery	773514681	LA8B.1	09.10.21	1	Absent or atretic right pulmonary artery	
280	260	09.10.77	Absent or atretic left pulmonary artery	211367900	LA8B.1	09.10.21	2	Absent or atretic left pulmonary artery	
281	261	09.10.37	Congenital central pulmonary arterial stenosis or hypoplasia proximal to hilar bifurcation	1061325287	LA8B.1	09.10.41	4	Congenital central pulmonary arterial stenosis or hypoplasia: proximal to hilar bifurcation	
282	262	09.10.38	Congenital peripheral pulmonary arterial stenosis or hypoplasia at or beyond hilar bifurcation	499211140	LA8B.1	09.10.41	5	Congenital peripheral pulmonary arterial stenosis or hypoplasia: at or beyond hilar bifurcation	

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Table 3. (*Continued*)

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283	263	09.10.30	Congenitally discontinuous, non-confluent right and left pulmonary arteries	620586359	LA8B.1	09.10.41	6	Congenitally discontinuous (non-confluent) right and left pulmonary arteries	Congenitally discontinuous (non-confluent) right and left pulmonary arteries
284	264	09.09.08	Pulmonary artery origin from ascending aorta	731364546	LA8B.1	09.10.41	7	Pulmonary artery origin from ascending aorta	
285	265	09.09.03	Right pulmonary artery from ascending aorta	2122410634	LA8B.1	09.09.08	1	Right pulmonary artery from ascending aorta	
286	266	09.09.05	Left pulmonary artery from ascending aorta	1284030093	LA8B.1	09.09.08	2	Left pulmonary artery from ascending aorta	
287	267	09.09.11	Pulmonary artery from arterial duct	16177108	LA8B.1	09.10.41	8	Pulmonary artery from arterial duct (ductus arteriosus)	Pulmonary artery ductus arteriosus
288	268	09.09.02	Right pulmonary artery from arterial duct	388422478	LA8B.1	09.09.11	1	Right pulmonary artery from arterial duct (ductus arteriosus)	Right pulmonary artery from ductus arteriosus
289	269	09.09.04	Left pulmonary artery from arterial duct	900008233	LA8B.1	09.09.11	2	Left pulmonary artery from arterial duct (ductus arteriosus)	Left pulmonary artery from ductus arteriosus
290	270	07.09.34	Congenital anomaly of aorta or its branches	1509021958	LA8B.2	09.04.28	3	Congenital anomaly of aorta and-or its branches	
291	271	09.16.06	Congenital anomaly of ascending aorta	1073599518	LA8B.2Y	07.09.34	1	Congenital anomaly of ascending aorta	
292	272	09.16.02	Hypoplasia of ascending aortic	1601805840	LA8B.2Y	09.16.06	1	Ascending aortic hypoplasia	
293	273	09.16.19	Congenital ascending aortic aneurysm or dilation	1381321493	LA8B.2Y	09.16.06	2	Congenital ascending aortic dilation or aneurysm; Congenital ascending aorta aneurysm or dilation	
294	274	09.28.10	Congenital anomaly of aortic arch	2071071585	LA8B.2Y	07.09.34	2	Congenital anomaly of aortic arch	
295	275	09.29.11	Hypoplasia of aortic arch	2084361271	LA8B.2Y	09.28.10	1	Aortic arch hypoplasia	
296	277	09.29.31	Interrupted aortic arch	1769930414	LA8B.22	09.28.10	2	Interrupted aortic arch	
297	278	09.29.32	Interrupted aortic arch distal to subclavian artery, type A	2023865940	LA8B.22	09.29.31	1	Interrupted aortic arch: distal to subclavian artery (type A)	

Table 3. (*Continued*)

298	279	09.29.33	Interrupted aortic arch between subclavian and common carotid arteries, type B	1512205361	LA8B.22	09.29.31	2	Interrupted aortic arch: between subclavian and common carotid arteries (type B)
299	280	09.29.34	Interrupted aortic arch between carotid arteries, type C	1840336207	LA8B.22	09.29.31	3	Interrupted aortic arch: between carotid arteries (type C)
300	281	09.28.15	Right aortic arch	769265824	LA8B.2Y	09.28.10	3	Right aortic arch
301	282	09.28.22	Left aortic arch	1624106993	LA8B.2Y	09.28.10	4	Left aortic arch
302	283	09.28.06	Cervical aortic arch	1466691538	LA8B.2Y	09.28.10	5	Cervical aortic arch
303	X^+	09.30.22	Aortic diverticulum of Kommerell	300462821	LA8B.2Y	09.28.10	6	
304	Y^+	09.28.08	Persistent fifth aortic arch	82536098	LA8B.2Y	09.28.10	7	
305	276	09.29.01	Coarctation of aorta	1524185114	LA8B.21	07.09.34	3	Coarctation of aorta
306	U^+	09.29.02	Preductal coarctation of aorta	1614972736	LA8B.21	09.29.01	1	
307	V^+	09.29.04	Postductal coarctation of aorta	1818445124	LA8B.21	09.29.01	2	
308	W^+	09.29.03	Juxtaductal (paraductal) coarctation of aorta	Awaiting ICD entity number	LA8B.21	09.29.01	3	
309	284	09.30.17	Congenital anomaly of aortic arch branch	1465829007	LA8B.24	07.09.34	4	Congenital anomaly of aortic arch branch
310	285	09.30.02	Aberrant origin of right subclavian artery	1753426940	LA8B.24	09.30.17	1	Aberrant origin of right subclavian artery
311	286	09.30.04	Aberrant origin of left subclavian artery	1980670431	LA8B.24	09.30.17	2	Aberrant origin of left subclavian artery
312	287	09.30.16	Isolation of an aortic arch branch	2051093194	LA8B.24	09.30.17	3	Isolation of an aortic arch branch
313	Z^+	09.30.11	Isolation of innominate artery	8058820	LA8B.24	09.30.16	1	
314	AA^+	09.30.14	Isolation of left subclavian artery	2066112447	LA8B.24	09.30.16	2	
315	AB^+	09.30.15	Isolation of right subclavian artery	77867676722	LA8B.24	09.30.16	3	
316	AC^+	09.30.12	Isolation of left common carotid artery	1612416791	LA8B.24	09.30.16	4	
317	AD^+	09.30.13	Isolation of right common carotid artery	1060908708	LA8B.24	09.30.16	5	
318	AE^+	09.30.34	Aberrant origin of innominate artery	224546005	LA8B.24	09.30.17	4	

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Table 3. (*Continued*)

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319	AF+	09.30.31	Common origin of the innominate artery and left common carotid artery	1476720756	LA8B.24	09.30.17	5		
320	AG+	09.30.28	Separate origins of internal and external carotid arteries	34296271	LA8B.24	09.30.17	6		
321	288	09.28.47	Congenital anomaly of descending thoracic or abdominal aorta	Awaiting ICD entity number	LA8B.23	07.09.34	5	Congenital anomaly of descending thoracic and/or abdominal aorta	
322	289	09.29.44	Descending thoracic or abdominal aortic coarctation	480830042	LA8B.23	09.28.47	1	Descending thoracic or abdominal aortic coarctation	
323	AH+	09.29.05	Coarctation of the descending thoracic aorta	1167364661	LA8B.23	09.29.44	1		
324	AI+	09.29.06	Coarctation of the abdominal aorta	566051188	LA8B.23	09.29.44	2		
325	290	09.31.40	Tracheo-oesophageal compressive syndrome	108967698	LA8B.3	09.04.28	4	Tracheo-esophageal compressive syndrome	
326	291	09.30.23	Innominate artery compression syndrome	837654124	LA8B.3	09.31.40	1	Innominate artery compression syndrome	
327	AJ+	09.30.27	Retro-oesophageal origin of aberrant innominate artery	1848026200	LA8B.3	09.30.23	1		
328	292	09.31.00	Vascular Ring	1864923481	LA8B.Y	09.04.28	5	Vascular Ring	
329	293	09.28.09	Double aortic arch	960436403	LA8B.Y	09.31.00	1	Double aortic arch	
330	294	09.31.35	Vascular ring of right aortic arch and left arterial duct or ligament	1921699903	LA8B.Y	09.31.00	2	Vascular ring of right aortic arch and left arterial duct or ligament	
331	295	09.31.34	Vascular ring of left aortic arch and right arterial duct or ligament	1256670334	LA8B.Y	09.31.00	3	Vascular ring of left aortic arch and right arterial duct or ligament	
332	296	09.09.06	Anomalous origin of left pulmonary artery from right pulmonary artery	1038861606	LA8B.Y	09.04.28	6	Anomalous origin of left pulmonary artery from right pulmonary artery (pulmonary arterial sling)	Pulmonary arterial sling
333	297	09.27.05	Congenital arterial duct anomaly	429811909	LA8B.Y	09.04.28	7	Congenital arterial duct (ductus arteriosus) anomaly	Congenital ductus arteriosus anomaly
334	298	09.27.21	Patent arterial duct	1262462321	LA8B.4	09.27.05	1	Patent arterial duct (ductus arteriosus)	Patent arterial duct
335	AK+	09.27.03	Absent arterial duct	316856895	LA8B.Y	09.27.05	2		

Table 3. (*Continued*)

336	AL+	09.27.04	Congenital aneurysm of arterial duct	1307324770	LA8B.Y	09.27.05	3		
337	AM+	09.27.41	Anomalous origin of arterial duct	1488672807	LA8B.Y	09.27.05	4		
338	AN+	09.27.82	Anomalous origin of arterial ligament	172834801	LA8B.Y	09.27.05	5		
339	AO+	14.10.51	Fetal arterial duct narrowing-closure	1802063004	LA8B.Y	09.27.05	6		
340	299	09.08.18	Systemic-to-pulmonary collateral arteries	807948372	LA8B.Y	09.04.28	8	Systemic-to-pulmonary collateral artery(ies)	Systemic-to-pulmonary collateral artery(ies)
341	300	09.46.03	Congenital anomaly of coronary artery	902783759	LA8C	01.01.59	10	Congenital anomaly of coronary arteries	
342	301	09.41.01	Anomalous origin of coronary artery from pulmonary arterial tree	1862930314	LA8C.0	09.46.03	1	Anomalous origin of coronary artery from pulmonary arterial tree	
343	302	09.41.03	Anomalous origin of left coronary artery from pulmonary artery	12665074	LA8C.0	09.41.01	1	Anomalous origin of left coronary artery from pulmonary artery	
344	303	09.42.00	Anomalous aortic origin or course of coronary artery	624468809	LA8C.1	09.46.03	2	Anomalous aortic origin or course of coronary artery	
345	304	09.42.21	Anomalous aortic origin of coronary artery with ventriculo-arterial concordance	238058271	LA8C.1	09.42.00	1	Anomalous aortic origin of coronary artery with ventriculo-arterial concordance	
346	305	09.46.26	Right coronary artery from left aortic sinus with ventriculo-arterial concordance	912125792	LA8C.1	09.42.21	1	Right coronary artery from left aortic sinus with ventriculo-arterial concordance	
347	306	09.46.21	Left coronary artery from right aortic sinus with ventriculo-arterial concordance	350108987	LA8C.1	09.42.21	2	Left coronary artery from right aortic sinus with ventriculo-arterial concordance	
348	307	09.43.04	Anterior descending from right coronary artery across right ventricular outflow tract	592256069	LA8C.1	09.42.00	2	Anterior interventricular (anterior descending) from right coronary artery across right ventricular outflow tract	Anterior interventricular from right coronary artery across right ventricular outflow tract
349	308	09.43.05	Intramural proximal coronary arterial course	1908546533	LA8C.1	09.42.00	3	Intramural proximal coronary arterial course	
350	309	09.43.13	Single coronary artery supplying all of heart	1238584494	LA8C.1	09.42.00	4	Single coronary supplying all of heart	

(*Continued*)

Table 3. (*Continued*)

ICD-11 New Row Number or Letter	ICD-11 Old Row Number or New Letter	IPCCC code	ICD-11 Congenital Cardiac term	ICD-11 Foundation entity number	ICD-11 MMS code or ICD-11 MMS crossmap	Parent	Sibling order	ICD-11 Congenital Cardiac term published in 2017[105] (terms in blue cells have been changed)	ICD-11 Congenital Cardiac term components published in 2017[105] and now moved to Synonyms
351	310	09.43.12	Myocardial bridging of coronary artery	184231124	LA8C.Y	09.46.03	3	Myocardial bridging of coronary artery	
352	311	09.44.05	Congenital coronary arterial orifice stenosis	1614456856	LA8C.Y	09.46.03	4	Congenital coronary arterial orifice stenosis	
353	312	09.44.19	Congenital coronary arterial orifice atresia	2118151636	LA8C.Y	09.46.03	5	Congenital coronary arterial orifice atresia	
354	313	09.45.16	Congenital coronary arterial fistula	1580310858	LA8C.2	09.46.03	6	Congenital coronary arterial fistula(s)	Congenital coronary arterial fistula(s)
355	314	09.45.10	Congenital coronary arterial fistula to right ventricle	955445461	LA8C.2	09.45.16	1	Congenital coronary arterial fistula to right ventricle	
356	315	09.45.22	Congenital coronary arterial fistula to left ventricle	98410917	LA8C.2	09.45.16	2	Congenital coronary arterial fistula to left ventricle	
357	316	09.46.14	Congenital coronary arterial aneurysm	1376805686	LA8C.Y	09.46.03	7	Congenital coronary arterial aneurysm(s)	Congenital coronary arterial aneurysm(s)
358	AP^{+}	09.42.09	Accessory coronary artery	1083781657	LA8C.Y	09.46.03	8		
359	AQ^{+}	09.46.44	Congenital absence of coronary artery	617293420	LA8C.Y	09.46.03	9		
360	AR^{+}	09.46.19	Coronary arterial hypoplasia	1156062809	LA8C.Y	09.46.03	10		
361	317	10.01.05	Congenital pericardial anomaly	1188459532	LA8D	01.01.59	11	Congenital pericardial anomaly	
362	AS^{+}	10.01.02	Complete agenesis of pericardium	1462683633	LA8D	10.01.05	1		
363	AT^{+}	10.01.01	Partial agenesis of pericardium	1742434564	LA8D	10.01.05	2		
364	AU^{+}	10.01.03	Pleuropericardial cyst	989128703	LA8D	10.01.05	3		
365	AV^{+}	10.03.53	Congenital cardiac tumour	awaiting ICD number.	LA8Y	01.01.59	12		
366	318***	09.19.05	Pulmonary arteriovenous fistula	913404991	LA90.5	01.01.59	13	Pulmonary arteriovenous fistula	
367	AW^{+}	02.02.03	Bifid apex of heart	1437052733	LA8Y	01.01.59	4		

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