

The International Working Group for Mapping and Coding of Nomenclatures for Paediatric and Congenital Heart Disease

Update from The International Working Group for Mapping and Coding of Nomenclatures for Paediatric and Congenital Heart Disease

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THE FIRST INTERNATIONAL SUMMIT ON Nomenclature for Congenital Heart Disease was held on 27 May, 2001, in Toronto, Canada, just prior to the 3rd World Congress of Pediatric Cardiology and Cardiac Surgery. At the Summit, enthusiastic and virtually unanimous support was expressed for the development of a unified system for describing congenital cardiac malformations, as well as the related procedures used to palliate or correct them. At the meeting of the International Nomenclature Committee for Paediatric and Congenital Heart Disease that immediately followed the Summit, a working group was appointed and given the

following tasks:

- Finalise the crossmap of the nomenclature “Short Lists” of the European Paediatric Cardiac Code¹ and the International Congenital Heart Surgery Nomenclature and Database Project,² both published in 2000.
- Crossmap the “Long Lists” and develop a single “super-tree” of nomenclature, to be called the International Paediatric and Congenital Cardiac Code, by the time of the next World Congress in Buenos Aires, Argentina.
- Report back to the International Committee to diffuse information about progress.

Mid-way through the time allotted to complete these tasks, therefore, we present a brief update on the progress of the working group to date. For additional details, we refer the reader to a full report outlining the background and events leading to the founding of the International Nomenclature Project for Paediatric and Congenital Heart Disease (Fig. 1), published in *Cardiology in the Young* in 2002.³

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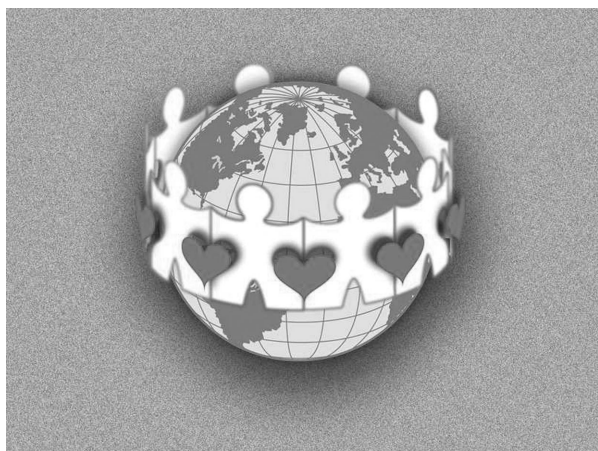


Figure 1.
Logo for the International Nomenclature Project for Paediatric and Congenital Heart Disease, presented at the First International Summit on Nomenclature for Congenital Heart Disease, May 27, 2001.

The Nomenclature Working Group

Six members were originally appointed to become the International Working Group for Mapping and Coding of Nomenclatures for Paediatric and Congenital Heart Disease. They were Vera D. Aiello, from Sao Paulo, Brazil; Marie J. Béland, from Montreal, Canada; Steven D. Colan, from Boston, USA; Rodney C. G. Franklin, from London, UK; Jeffrey P. Jacobs, from St. Petersburg, USA; and Christo I. Tchervenkov, from Montreal, Canada. To enhance international representation, and balance the number of surgical versus non-surgical members, an additional six individuals were added to the group. They were: J. William Gaynor, from Philadelphia, USA; Otto N. Krogmann, from Duisburg, Germany; Hiromi Kurosawa, from Tokyo, Japan; Bohdan Maruszewski, from Warsaw, Poland; Giovanni Stellin, from Padova, Italy; and Paul M. Weinberg, from Philadelphia, USA.

Since its inception, the Working Group has met four times, with each meeting lasting from three to five days. The first three meetings were held in Canada, while the fourth was held in Brazil. We have planned three more meetings to be held prior to the 4th World Congress of Pediatric Cardiology and Cardiac Surgery in Argentina, which itself will be held in September, 2005 (Table 1).

Crossmapping the Nomenclature Lists and building the International Paediatric and Congenital Cardiac Code

Crossmapping the "Short Lists"

The "Short Lists" of the European Paediatric Cardiac Code¹ and the International Congenital Heart Surgery

Table 1. Meetings of the Nomenclature Working Group.

Location	Date
Montreal, Canada	May 9–13, 2002
Montreal, Canada	January 17–19, 2003
Lake Louise, Canada	April 2–6, 2003
Recife, Brazil	December 10–14, 2003
Warsaw, Poland	Planned for July 2004
Montreal, Canada	Planned for December 2004
Saint Petersburg, USA	Planned for May 2005
Buenos Aires, Argentina	Planned for September 2005

Nomenclature and Database Project² were bidirectionally crossmapped by the Executive of the Nomenclature Working Group, made up of Marie J. Béland, Rodney C. G. Franklin, Jeffrey P. Jacobs, and Christo I. Tchervenkov. This work resulted in the publication of the two-way crossmap in 2002.^{4,5}

The United Kingdom Central Cardiac Audit Database collects congenital heart surgery outcome data using the Short List established by the Association of European Paediatric Cardiology, also known as the European Paediatric Cardiac Code. The European Association for Cardio-thoracic Surgery Database collects data relating to the outcomes of surgery for congenital cardiac malformations using the Short List established by the International Congenital Heart Surgery Nomenclature and Database Project. The bidirectional crossmap between these two short lists will permit centres or surgeons coding in either system to submit data to both registries with one institutional database. Efforts are currently underway by the United Kingdom Central Cardiac Audit Database to facilitate this process, which would represent the first multi-institutional clinical application of the crossmaps.

Crossmapping the "Long Lists"

The crossmapping of the "Long Lists" is currently being carried out by the Nomenclature Working Group at semi-annual meetings. Prior to each meeting, the executive members of the group prepare an agenda listing the lesions to be reviewed. The lesions are selected from the table of contents of the International Congenital Heart Surgery Nomenclature and Database Project.² A member of the Working Group is assigned a lesion to review, and is expected to present the pertinent related nomenclatures, highlighting synonymous terms, and/or any controversies that may exist among the schemes for classification already extant. After each presentation to the group, the entries from the long list of the International Congenital Heart Surgery Nomenclature and Database Project are crossmapped one by one to equivalent terms from the European Paediatric Cardiac Code.

If there is no equivalent term, one is added, along with a numeric code conforming to the method of numerical codification of the European Paediatric Cardiac Code. The process of crossmapping is then carried out in the reverse direction, such that each entry from the long list of the European Paediatric Cardiac Code is reviewed, and its equivalency is sought on the list of the International Congenital Heart Surgery Nomenclature and Database Project. Omissions to the latter list are then added.

During the process of bidirectional crossmapping between the "Long Lists", additional entries may be suggested from the collective expertise of the members of the Nomenclature Working Group, including terms that appear in the Nomenclature Tree of the Canadian Pediatric Cardiology Association, as yet unpublished, and the Fyler codes from Boston, also unpublished.

Each meeting of the Working Group begins with a session for the detailed review of lesions discussed at previous meetings. This half-day session is entitled "Omissions and Errors". Its purpose is to have a second look at lesions already crossmapped, so as to ensure the completeness and accuracy of the process. Lesions and related procedures discussed during the first four meetings of the group are shown in Table 2. Through this elaborate and collaborative effort, we are meticulously constructing the International Paediatric and Congenital Cardiac Code.

The International Classification of Diseases of the World Health Organization

Paediatric cardiologists and cardiac surgeons have long considered that the nomenclature and codes of the *International Statistical Classification of Diseases and Related Health Problems, Ninth and Tenth Revisions*, known as "ICD-9" and "ICD-10", provide insufficient detail for reporting lesions pertinent to their practice. In addition, cardiac procedures are not covered in the official versions provided by the World Health Organization. Notwithstanding these limitations, hospital administrators in many countries continue to require that these International Classifications be used. To reconcile the needs of paediatric cardiologists and cardiac surgeons with those of administrators, the International Paediatric and Congenital Cardiac Code will be crossmapped to both the Ninth and Tenth Revisions.

The World Health Organization has been apprised of the work of our Group. The Update Reference Committee of the World Health Organization recently consulted the group to clarify the classification of Eisenmenger syndrome and Eisenmenger defect for their deliberations in October, 2003. We anticipate still further collaboration with the World Health

Table 2. Lesions and related procedures reviewed to date and to be reviewed.

First Four Meetings

1. Interatrial Communications
2. Ventricular Septal Defects
3. Atrioventricular Septal Defects
4. Aortopulmonary Windows
5. Common Arterial Trunk (Truncus Arteriosus)
6. Pulmonary Venous Anomalies
7. Systemic Venous Anomalies
8. Tetralogy of Fallot
9. Pulmonary Atresia with Ventricular Septal Defect
10. Ebstein's Malformation and Other Tricuspid Valvar Anomalies
11. Aortic Valvar Disease
12. Mitral Valvar Anomalies
13. Aortic Aneurysm and Dissection
14. Hypoplastic Left Heart Syndrome
15. Patency of the Arterial Duct

Next Three Meetings

1. Right Ventricular Outflow Tract Obstruction with an Intact Ventricular Septum
2. Functionally Univentricular Heart
3. Concordant Atrioventricular Connections with Discordant Ventriculo-arterial Connections (Transposition of the Great Arteries)
4. Congenitally Corrected Transposition
5. Double Outlet Right Ventricle
6. Double Outlet Left Ventricle
7. Aorto-ventricular Tunnel
8. Coarctation of the Aorta, Interrupted Aortic Arch
9. Vascular Rings
10. Cardiac Malpositions

Future Meetings After Fourth World Congress

1. Cardiomyopathies and End-stage Congenital Heart Disease
2. Diseases of the Pericardium
3. Coronary Arterial Abnormalities
4. Arrhythmias
5. Interventional Procedures
6. End-stage Lung Disease
7. Cardiac Tumours
8. Fetal Cardiology
9. Adults with Congenital Heart Disease
10. Paediatric Critical Cardiac Care

Organization, and hope that this will result in the dissemination by the World Health Organization of lists of names that are more practical for the fields of paediatric and congenital cardiology and cardiac surgery.

Goals of the Working Group for 2004 and 2005

Completion of the crossmap of congenital heart disease and related procedures

We expect to have crossmapped, in bidirectional fashion, the remaining major cardiac lesions by September, 2005. Several challenging lesions have yet to be discussed, including "functionally single ventricle" and "double outlet right ventricle" (Table 2).

Ensuring global understanding of, and support for, The International Nomenclature Project

It is planned that, during 2004, all of the main national and continental organizations with responsibilities for children and adults with congenital heart disease will be contacted by the International Nomenclature Committee, informing them formally of our ongoing project. It is hoped that the aims of the International Nomenclature Committee, with its Working Group, will be both endorsed and supported, and that this endorsement will then be cascaded down to the individual members of these various bodies.

Obtaining further funding

Funding for the first four meetings of the Nomenclature Working Group was generously provided, in part, by the Canadian Institutes of Health Research, the Heart and Stroke Foundation of Canada, and the Montreal Children's Hospital Foundation. Additional funding will be sought for the support of future meetings.

Cooperation and integration with other international projects relating to nomenclature

Our group is aware that the *Systematized Nomenclature of Medicine Clinical Terms*, also known as the SNOMED CT project, which details terms with a numerical coding system from across all medical specialties, has major sections on paediatric cardiology and cardiac surgery. This system was originally developed by the College of American Pathologists,⁶ and has more recently amalgamated with the Clinical Terms Version 3 developed in the United Kingdom, and formally known as the Read coding system, with an aim to become an integral part of the electronic patient record on both sides of the Atlantic.⁷ The Working Group plans to link with the further development of this important system so as to have the final International Paediatric and Congenital Cardiac Code integrated into the Systematized Nomenclature of Medicine Clinical Terms.

Organization of the Second Summit

The Second International Summit on Nomenclature for Paediatric and Congenital Heart Disease will be organized by the Working Group, and will be held during the 4th World Congress of Pediatric Cardiology and Cardiac Surgery in Buenos Aires, Argentina. This will take place from September 18–22, 2005. It is anticipated that this will be the launch of the International Paediatric and Congenital Cardiac Code, a global venture to harmonize the various

disparate systems for coding and nomenclature that exist throughout the world. We expect that this will enable transcontinental cooperative studies further to advance the fields of paediatric cardiology and cardiac surgery.

The Future After the 4th World Congress

The Working Group will be planning the next phase of its work after the next meeting of the International Committee, which will take place during the Second International Summit, planned for the World Congress in September, 2005. Interventional procedures, arrhythmias, cardiac tumours, cardiomyopathies, fetal congenital cardiac disease, and adults with congenital heart disease, are but some of the topics remaining to be reviewed after the World Congress.

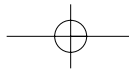
Provision must be made for the renewal of the membership of the Nomenclature Working Group, and for the upkeep and revision of the International Paediatric and Congenital Cardiac Code. The "super-tree" of nomenclature for paediatric and congenital cardiac disease not only needs to be completed, but also a solid foundation needs to be established for continued revitalization and growth of the tree as new concepts, discoveries, and developments evolve. Much work remains to be done, and continued support of the project by the paediatric cardiologic and cardiovascular communities is much appreciated. Further concerted efforts will continue to ensure the successful completion and adoption of the International Paediatric and Congenital Cardiac Code across the world.

Acknowledgments

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References

1. Association for European Paediatric Cardiology. The European Paediatric Cardiac Code. *Cardiol Young* 2000; 10 (Suppl 1): 1–146.
2. Mavroudis C, Jacobs JP (eds.). The International Congenital Heart Surgery Nomenclature and Database Project. *Ann Thorac Surg* 2000; 69 (S1): 1–372.
3. Béland MJ, Jacobs JP, Tchervenkov CI, Franklin RCG. The International Nomenclature Project for Paediatric and Congenital Heart Disease: Report from the Executive of The International Working Group for Mapping and Coding of Nomenclatures for Paediatric and Congenital Heart Disease. *Cardiol Young* 2002; 12: 425–430.



4. Franklin RCG, Jacobs JP, Tchervenkov CI, Béland MJ. Report from the Executive of The International Working Group for Mapping and Coding of Nomenclatures for Paediatric and Congenital Heart Disease: Bidirectional Crossmap of the Short Lists of the European Paediatric Cardiac Code and the International Congenital Heart Surgery Nomenclature and Database Project. *Cardiol Young* 2002; 12 (Suppl 2): 18–22.
5. Franklin RCG, Jacobs JP, Tchervenkov CI, Béland MJ. The International Nomenclature Project for Congenital Heart Disease: Bidirectional Crossmap of the Short Lists of the European Paediatric Cardiac Code and the International Congenital Heart Surgery Nomenclature and Database Project. *Cardiol Young* 2002; 12: 431–435.
6. College of American Pathologists; Systematized Nomenclature of Medicine, 2 vols, 2nd edn; Côté RA, editor-in-chief. College of American Pathologists, Skokie, Illinois, 1979.
7. Stearns MQ, Price C, Spackman KA, Wang AY. SNOMED clinical terms: overview of the development process and project status. *Proc AMIA Symp* 2001: 662–666.

