

## *The International Nomenclature Project for Congenital Heart Disease*

# Report from the Executive of The International Working Group for Mapping and Coding of Nomenclatures for Paediatric and Congenital Heart Disease

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IT IS ONLY IN THE LAST CENTURY THAT THE STUDY of congenital cardiac malformations began in earnest. Thus, as recently as 1936, the distinguished Canadian physician, Maude Abbott, published the *Atlas of Congenital Cardiac Disease*,<sup>1</sup> this representing the first formal system of classification for cardiac malformations. Subsequent to this initial venture, however, in an effort to establish individual hospital-specific databases, centres across the world have developed or adopted different schemes for classification of the diagnoses and procedures related to congenital cardiac disease. Unfortunately, the disparate systems thus evolved have rather served to make impracticable interinstitutional and international research into the epidemiology of congenital cardiac disease. In addition, while the demand for multicentric analysis of outcomes, and stratification of risk, for interventions in children with heart disease has increased, such evaluations cannot easily be performed without a uniform system of nomenclature.

### Efforts to bring uniformity to nomenclature for congenital cardiac disease

Recognizing the importance of having a uniform system for the nomenclature of congenital cardiac malformations, there have been several efforts since the 1970s to establish broad-based common lists.

The development of computers, and advances in the technology of communication, have greatly facilitated the task of standardization of entry of data and exchange of information. In the late 1990s, two major initiatives developed in parallel, one involving mainly European paediatric cardiologists and the other, chiefly paediatric cardiac surgeons from North America and Europe. The Association for European Paediatric Cardiology, including representation from the European Association for Cardiothoracic Surgery, published its European Paediatric Cardiac Codes in *Cardiology in the Young* as a supplement to volume 10 in January 2000.<sup>2</sup> On the surgical side, representatives of the Society of Thoracic Surgeons and the European Association for Cardiothoracic Surgery developed their own diagnosis and procedure list, the International Congenital Heart Surgery Nomenclature and Database Project, that was published in *The Annals of Thoracic Surgery* as a supplement to volume 69 in April 2000.<sup>3</sup> Both the systems included a Long List and a Short List, the Short Lists being composed of a condensed series of terms, with 434 entries on the surgical side, and 650 in the European Paediatric Cardiac Code. The Long Lists, on the other hand, consisted of many thousands of entries in both systems. Although there is a similar underlying aim of the two systems, namely to establish a standardised nomenclature for congenital cardiac disease, the overriding drive for the more surgical system was to provide a list which would be the basis for collection of surgical data, so as to enable multicentric and international analysis of surgical outcomes taking account of case-mix along with stratification of risk.

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Of note is that there are no numerical codes attached to the terms within the International Congenital Heart Surgery Nomenclature and Database Project.

### The International Nomenclature Project for Paediatric and Congenital Heart Disease

In recognition of the problematic situation of having two lists that were to be widely adopted, one mostly by paediatric cardiologists, the other mainly by paediatric cardiac surgeons, a meeting was called between representatives from the Association for European Paediatric Cardiology, the Society of Thoracic Surgeons, and the European Association for Cardiothoracic Surgery. This meeting was held on 6 October, 2000, in Frankfurt, Germany, at the European Congenital Heart Surgeons Foundation meeting prior to the 14th Annual Meeting of the European Association for Cardiothoracic Surgery. Without a collaborative effort between these groups, there was a real risk of invalidating multicentric trials due to confusion between the coding systems and potentially duplicate or inaccurate entries within institutions. It was agreed in Frankfurt that representatives of the societies should work in partnership, and produce a reconciliatory bidirectional map between the two systems. The feasibility of this would be established initially by creating a crossmap between the "Short Lists" of the two systems for presentation at the Third World Congress of Pediatric Cardiology and Cardiac Surgery in Toronto, Canada in May 2001. It was also resolved that a larger international nomenclature meeting should be organized and held at the time of the World Congress to discuss the possibility of establishing a common international system of nomenclature for congenital cardiac malformations. The International Nomenclature Project for Paediatric and Congenital Heart Disease was thus founded on October 6th, 2000 in Frankfurt, Germany, to be fostered by a newly formed International Nomenclature Committee for Paediatric and Congenital Heart Disease, chaired by Martin Elliott of London, England.

### The first International Summit on Nomenclature for Congenital Heart Disease

The first International Summit on Nomenclature for Congenital Heart Disease was held on 27 May, 2001, in Toronto, Canada, just prior to the Third World Congress of Pediatric Cardiology and Cardiac Surgery (Fig. 1). The meeting was organized by Marie J. Béland and Christo I. Tchervenkov from Montreal, Canada, and attended by over 250 people. Surgical associations and cardiologic societies from around the world were represented, including the societies



Figure 1.

*The logo of the International Nomenclature Project for Congenital Heart Disease, which was unveiled at the first International Summit on Nomenclature for Congenital Heart Disease, May 27th, 2001, Toronto, Canada.*

and associations producing the European Paediatric Cardiac Code and the International Congenital Heart Surgery Nomenclature and Database Project, as well as the American Association for Thoracic Surgery, the American College of Cardiology, the Asian Society of Cardiovascular Surgery, the Canadian Cardiovascular Society, the Cardiology Subsection of the American Academy of Pediatrics, the Congenital Heart Surgeons Society, and the Council on Cardiovascular Disease in the Young of the American Heart Association. Additional participants included representatives from Asia, Australia, and South America. The program included presentations from cardiologists and surgeons from North America and Europe (Table 1). At the meeting, the concept of crossmapping nomenclature systems was introduced by Jeffrey P. Jacobs, and that of a single "super-tree" of nomenclature by Marie J. Béland. An initial version of the crossmap between the two Short Lists had been created by Rodney C.G. Franklin, and was presented with a report of progress.

The summit was attended by the acknowledged experts in morphology, Robert H. Anderson from London, United Kingdom, and Richard Van Praagh from Boston, United States of America. Both were invited to comment, and both expressed their strong support for the development of a unified system for describing congenital cardiac malformations. In the panel discussions that followed the presentations to the summit, virtually unanimous support was expressed for the concept of a common international nomenclature tree.

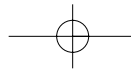


Table 1. Program of The International Summit on Nomenclature for Congenital Heart Disease held on May 27th, 2001 in Toronto, Canada.

<b>Moderators</b>	Christo I. Tchervenkov, Montreal, Canada Constantine Mavroudis, Chicago, USA
Welcome	Christo Tchervenkov, Montreal, Canada
Introduction and Aims of the International Nomenclature Committee	Martin Elliott, London, United Kingdom
The View of the European Association for Cardio-Thoracic Surgery,	Bohdan Maruszewski, Warsaw, Poland
The View of the Society of Thoracic Surgeons	Constantine Mavroudis, Chicago, USA
The View of the Association for European Paediatric Cardiology	Franco Stocker, Muri, Switzerland
The View of the American Society of Echocardiography	Steven Colan, Boston, USA
A Canadian Unifying View	Marie Béland, Montreal, Canada
Introduction to Mapping and Computer Based Mapping Solutions	Jeffrey Jacobs, St. Petersburg, USA
The Specifics of Short List Mapping	Rodney Franklin, London, United Kingdom
Practical Experience with Databases: The Congenital Heart Surgeons Society and The Toronto Hospital for Sick Children	William G. Williams, Toronto, Canada
Risk Stratification for Congenital Heart Disease: International Complexity Score Study	François Lacour-Gayet Hamburg, Germany
Related Issues and Future Plans	Panel
Concluding Remarks	Constantine Mavroudis, Chicago, USA

Table 2. Initial membership of The International Nomenclature Committee for Congenital Heart Disease.

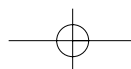
Martin J. Elliott – Chairman	London, United Kingdom	ECHSF, EACTS, AEPC
Christo I. Tchervenkov – Secretary	Montreal, Canada	STS, AATS, CHSS, CCS
Marie J. Béland – Treasurer	Montreal, Canada	Canadian Pediatric Cardiology Association, CCS
Vera D. Aiello	Sao Paulo, Brazil	Sociedade Brasileira de Cardiologia
Robert H Anderson	London, United Kingdom	AEPC
Steven D. Colan	Boston, USA	ACC, CVDY
Tjark Ebels	Groningen, The Netherlands	EACTS, ECHSF
Rodney C.G. Franklin	London, United Kingdom	AEPC
J. William Gaynor	Philadelphia, USA	STS, AATS
Jeffrey P. Jacobs	St. Petersburg, USA	STS
Kathy Jenkins	Boston, USA	AHA, CVDY
Christian Kreutzer	Buenos Aires, Argentina	South American surgeons
Otto N. Krogmann	Duisburg, Germany	AEPC
Hiromi Kurosawa	Tokyo, Japan	Japanese paediatric cardiologists and surgeons
Bohdan Maruszewski	Warsaw, Poland	EACTS, ECHDD (Director)
Constantine Mavroudis	Chicago, USA	STS (Chairman Database Committee), AATS, CHSS
Brian McCrindle	Toronto, Canada	Canadian Pediatric Cardiology Association
John Moore	Philadelphia, USA	ACC
Jack Rome	Philadelphia, USA	AHA, CVDY
David Sahn	Oregon, USA	ACC
Giovanni Stellin	Padova, Italy	EACTS
Franco Stocker	Muri, Switzerland	AEPC
Herbert E Ulmer	Heidelberg, Germany	AEPC
Carole Warnes	Rochester, USA	ACC
Paul Weinberg	Philadelphia, USA	AHA, CVDY
Jim Wilkinson	Melbourne	Australian Paediatric Cardiology

Abbreviations: AATS: American Association for Thoracic Surgery; ACC: American College of Cardiology; AEPC: Association for European Paediatric Cardiology; AHA: American Heart Association; CCS: Canadian Cardiovascular Society; CHSS: Congenital Heart Surgeons' Society; CVDY: Council on Cardiovascular Disease in the Young of the AHA; EACTS: European Association for Cardio-Thoracic Surgery; ECHDD: European Congenital Heart Defects Database; ECHSF: European Congenital Heart Surgeons Foundation; STS: Society of Thoracic Surgeons.

Note: Efforts are currently underway to increase involvement from paediatric cardiologists and surgeons from Africa, Asia, Australasia and South America. The Chairman and secretary would therefore welcome approaches from other societies globally who would be interested in having representation on this committee. The membership will be rationalised in the light of these approaches and the need for global representation, before it is finalised by the end of 2002.

The first meeting of the International Nomenclature Committee for Paediatric and Congenital Heart Disease was held immediately after the Summit (Table 2). Representatives from Asia, Europe, North

and South America were present for the deliberations. During the meeting, it was reiterated that the European Paediatric Cardiac Code has a more cardiological lean, while the International Congenital Heart



Surgery Nomenclature and Database Project favours surgical procedures and databases. The meeting ended by endorsing the view that the two coding hierarchies were complimentary, and should not be seen as competitive. To reconcile the two systems, a subcommittee entitled "The International Working Group for Mapping and Coding of Nomenclatures for Paediatric and Congenital Heart Disease" was formed, to have the abridged name of the Nomenclature Working Group, and given three mandates:

- To finalise the crossmapping of the two Short Lists of each system of nomenclature within the next year.
- To crossmap the "Long" Lists, and/or develop a single "super-tree" of nomenclature, by the time of the next World Congress in Buenos Aires, Argentina in 2005. The process was to be initiated by using numerical codes based on the six digit numbers of the European Paediatric Cardiac Code as its backbone.
- To report back regularly to the larger International Nomenclature Committee for Paediatric and Congenital Heart Disease, which in turn would communicate with associations for cardiology, cardiac surgery, and cardiovascular disease throughout the world about progress.

It must be emphasised that membership of the International Nomenclature Committee for Paediatric and Congenital Heart Disease has yet to be finalised, and that approaches would be welcome from other societies globally who might be interested in having representation on this committee.

### Crossmapping of the Short Lists

Since the establishment of the Nomenclature Working Group in Toronto in May 2001, finalization of the bidirectional crossmap of the two Short Lists was completed by the Executive members of the group: Rodney C.G. Franklin, Jeffrey P. Jacobs, Christo I. Tchervenkov, and Marie J. Béland. This work has resulted in the addition of the crossmapped terms of the Short List of the International Congenital Heart Surgery Nomenclature and Database Project that appear as the eighth column in the revised Short List of the European Paediatric Cardiac Code, published in the supplement to this issue of *Cardiology in the Young*.<sup>4</sup> The reverse mapping of the recently updated surgical Short List to the European Paediatric Cardiac Code Short List is also provided within the supplement.<sup>5</sup> Details of this process are given in the accompanying Supplement to this edition of *Cardiology in the Young*.<sup>6</sup>

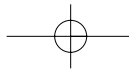
### Beyond crossmapping the Short Lists: The International Working Group for the Mapping and Coding of Nomenclatures for Paediatric and Congenital Heart Disease

After crossmapping the Short Lists of both the European Paediatric Cardiac Code and the International Congenital Heart Surgery Nomenclature and Database Project, the second and most important mandate of the Nomenclature Working Group was to crossmap the Long Lists of the two systems, and/or develop a single "super-tree" of nomenclature, by the time of the next World Congress in Buenos Aires, Argentina in 2005. The Nomenclature Working Group includes representation from paediatric cardiac surgery, paediatric cardiology and pathology, as well as members from Asia, Europe, North and South America (Table 3). The group convened in Montreal, Canada for its first formal meeting from May 9th to May 13th, 2002, to elaborate strategies to build an international unified nomenclature tree. It was agreed in Montreal that, over the next three years, the group would work toward the creation of a comprehensive and all-inclusive international system for nomenclature for paediatric and congenital cardiac disease using numerical codes based on the six digit numbers from the European Paediatric Cardiac Code as its backbone. This work would be accomplished through the process of bidirectional crossmapping of the European Paediatric Cardiac Code<sup>2</sup> and the International Congenital Heart Surgery Nomenclature and Database Project,<sup>3</sup> with additional mapping to the following lists, presented in alphabetical order: the Canadian Congenital Heart Codes, as yet unpublished; the Fyler codes developed in Boston by Donald C. Fyler, and also as yet unpublished; and the 9th and 10th revisions of the International Classification of Diseases, the latter provided by the World Health Organization.

Table 3. Members of the The International Working Group for Mapping of Nomenclatures for Paediatric and Congenital Cardiac Disease: the "Nomenclature Mapping Group".

Vera D. Aiello	Sao Paulo, Brazil
Marie J. Béland	Montreal, Canada* – Secretary-Treasurer
Steven D. Colan	Boston, USA
Rodney C.G. Franklin	London, United Kingdom*
J. William Gaynor	Philadelphia, USA
Jeffrey P. Jacobs	St. Petersburg, USA*
Bohdan Maruszewski	Warsaw, Poland
Giovanni Stellin	Padova, Italy
Christo I. Tchervenkov	Montreal, Canada* – Chairman
<i>Final Additional members (as of May, 2002)</i>	
Otto N. Krogmann	Duisburg, Germany
Hiromi Kurosawa	Tokyo, Japan
Paul Weinberg	Philadelphia, USA

\*Nomenclature Mapping Group Executive Committee Members.



After first establishing the structure of the group, along with its goals, and the methodology to achieve these goals, the work of "Long List" crossmapping was begun. A one-for-one process was established for the bidirectional mapping of the two Long Lists, with areas of deficiency in either listing being incorporated into the other, as well as newer terms with codes being created where necessary to enable the system to be fully comprehensive. Pertinent entries from the other lists, such as the Fyler codes from Boston, were integrated as needed. Synonymous terms were identified and attributed a single six digit number derived from the European Paediatric Cardiac Code. The Nomenclature Working Group did not undertake to judge which terms were the most scientifically accurate in describing a phenotype. Instead, it strove to identify terms that describe the same entity, and to group these under one code number. In the future, given such a numerical backbone and being fully comprehensive, other coding systems or lists of names will be able similarly, whatever the language, to crossmap terms to the international "master" tree, the International Paediatric Cardiac Code, being built by the Nomenclature Working Group.

At this first meeting in Montreal, five cardiac malformations were examined and successfully mapped bidirectionally. These were atrial septal defects; ventricular septal defects; atrioventricular septal defects; anomalies of systemic venous return; and common arterial trunk. The fruit of this initial work is already included in the revised version of the European Paediatric Cardiac Code, published in the Supplement of this issue of *Cardiology in the Young*.<sup>7</sup> At least five more meetings are planned over the next three years to complete the review of all paediatric and congenital cardiac disease, including related disorders and procedures. Regular reports on progress will be forwarded to the larger International Nomenclature Committee for Paediatric and Congenital Heart Disease. The International Nomenclature Committee, in turn, will circulate updates to all affiliated associations and societies on a regular basis.

### The future

The Nomenclature Working Group has been given a little more than three years to establish a nomenclature "super-tree" to be ready for international ratification by the time of the next World Congress of Pediatric Cardiology and Cardiac Surgery in Buenos Aires in 2005. In completing this task, the goals of the International Nomenclature Committee for Paediatric and Congenital Heart Disease will be accomplished, and a comprehensive and definitive International Paediatric Cardiac Code will be created. Research

will be facilitated across disciplines and internationally thanks to global collaboration. Once established, this all-inclusive system for nomenclature will facilitate international stratification of risk, and scoring of complexity, in cardiovascular disease continuing far beyond 2005. Institutional morbidity and mortality can then be reviewed, and compared to the relevant international gold standard. This accomplishment would continue to place paediatric cardiology and paediatric cardiac surgery at the forefront of medical practice in terms of establishing and maintaining valid international standards of best care and outcome for patients of all ages with cardiac disease.

### Websites

The Short Lists of the European Paediatric Cardiac Code (current version) and the International Congenital Heart Surgery Nomenclature and Database Project (version as published in 2000<sup>3</sup>) are available on the following sites, respectively:

[www.aepc.org](http://www.aepc.org),  
[www.pediatric.ecsur.org](http://www.pediatric.ecsur.org), and  
[www.sts.org](http://www.sts.org).

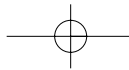
### Acknowledgements

The authors acknowledge the tremendous contribution of all individuals who have worked on the establishment of the European Paediatric Cardiac Code and the International Congenital Heart Surgery Nomenclature and Database Project. The Nomenclature Working Group wishes to thank Robert H. Anderson and Richard Van Praagh for their valued contributions and encouragement with the process of crossmapping the Long Lists.

The International Summit on Nomenclature for Congenital Heart Disease was made possible in part by the generous support of the Montreal Children's Hospital Foundation, and the organizing committee of the Third World Congress of Pediatric Cardiology and Cardiac Surgery. The authors also gratefully acknowledge the part sponsorship of the meetings of the Nomenclature Working Group by the Institute of Circulatory and Respiratory Health of the Canadian Institutes of Health Research. The logo for the International Nomenclature Project for Congenital Heart Disease was designed and produced by Marie J. Béland, Stephen J. Korkola and Christo I. Tchervenkov.

### References

1. Abbott ME. Atlas of Congenital Cardiac Disease. New York: The American Heart Association, 1936.
2. Association for European Paediatric Cardiology. The European Paediatric Cardiac Code. *Cardiol Young* 2000; 10 (Suppl. 1): 1-146.



3. Congenital Heart Surgery Nomenclature and Database Project. *Ann Thorac Surg* 2000; 69(4) (Suppl.): S1-S372.
4. Short List of European Paediatric Cardiac Code with crossmap to the Short List of International Congenital Heart Surgery Nomenclature and Database Project. *Cardiol Young* 2002; 12 (Suppl. 2).
5. Short List of the International Congenital Heart Surgery Nomenclature and Database Project with crossmap to the Short List of the European Paediatric Cardiac Code. *Cardiol Young* 2002; 12 (Suppl. 2).
6. Franklin RCG, Jacobs JP, Tchervenkov CI, Béland M. Bidirectional Crossmap of the Short Lists of the European Paediatric Cardiac Code and the International Congenital Heart Surgery Nomenclature and Database Project. Report from the Executive of The International Working Group for Mapping and Coding of Nomenclatures for Paediatric and Congenital Heart Disease. *Cardiol Young* 2002; (Suppl. 2).
7. Association for European Paediatric Cardiology. The European Paediatric Cardiac Code. The Long List with crossmaps to the 9th and 10th revisions of the International Classification of Disease and to the Short List of the European Paediatric Cardiac Code. *Cardiol Young* 2002; 12 (Suppl. 2).

