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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

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On 6 October, 2000, a meeting of representatives from the Association for European Paediatric Cardiology, the Society of Thoracic Surgeons, and the European Association for Cardiothoracic Surgery, took place in Frankfurt, Germany to discuss the publications earlier that year of two separate systems of nomenclature for paediatric and congenital heart disease: the European Paediatric Cardiac Code[1] and the International Congenital Heart Surgery Nomenclature and Database Project.[2] It was agreed at this meeting that the Short Lists of both systems should be mapped to each other in a first attempt to gravitate toward a single system for describing cardiac defects and procedures related to the heart. The need for this mapping, the historical background of the two parallel nomenclature systems and the later ratification of the mapping process by the first International Summit on Nomenclature for Congenital Heart Disease on 27 May, 2001, in Toronto, Canada, are discussed in the current issue of *Cardiology in the Young*.[3] Immediately following the Toronto Summit, the International Nomenclature Committee for Paediatric and Congenital Heart Disease met together, and created the International Working Group for Mapping and Coding of Nomenclatures for Paediatric and Congenital Heart Disease. This Nomenclature Working Group was mandated, within the next year, to finalise the crossmapping of the two Short Lists of each nomenclature system. Thereafter, the bidirectional crossmap of the two Short Lists was completed by the Executive committee of the group, namely Rodney C.G. Franklin, Jeffrey P. Jacobs, Christo I. Tchervenkov, and Marie J. Béland.

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Crossmapping of the Short Lists

The mapping process began by using the preliminary bidirectional crossmap provided by one of the Executive (RCGF) as a basis for detailed discussion just prior to, and during, the year following the Toronto meeting. The Short List of the International Congenital Heart Surgery Nomenclature and Database Project was then scrupulously examined by another member of the Executive (MJB) with respect to the corresponding terms in the European Paediatric Cardiac Code. A series of discussion points were raised during the Autumn of 2001, with a series of 11 instalments sent electronically to the other members of the Executive. After several discussions using electronic mail with proposed modifications to the two Short Lists, the mapping was brought to near completion during four, three-hour long, telephone conference calls between the Executive members. The crossmap between the two systems was finalised at the commencement of the Montreal meeting of the International Working Group for Mapping and Coding of Nomenclatures for Paediatric and Congenital Heart Disease in May 2002.

During this process, the crossmapped terms of the Short List of the International Congenital Heart

Surgery Nomenclature and Database Project was added as an eighth column to the revised Short List of the European Paediatric Cardiac Code, now published in this supplement of *Cardiology in the Young*.[4] In the reverse mapping, the term in the Short List of the European Paediatric Cardiac Code, and its six digit numerical code, appear as the second and third columns of the recently updated spreadsheet listing of the Surgical Nomenclature Short List within this supplement.[5] The following four columns in the latter Short List consist of the appropriate codes of the 9th and 10th revisions of the International Classification of Diseases, as described in the accompanying article in this supplement.[6]

To ensure that each diagnosis and procedure in one Short List could be mapped to an equivalent in the other, it was clear from the outset of the crossmapping process that it would be necessary to create new terms for each list. Also, when indicated, certain terms would need to be promoted from the Long List of each nomenclature system to their respective Short List. Although initially a full bidirectional map was envisaged, such that each item on each Short List would have its equivalent in the other, it soon became clear that the differing remits of the organisations involved would render this more difficult for the Short Lists. The Short List of the Congenital Heart Surgery Nomenclature and Database Project was designed as a minimal dataset for specific surgical needs. Furthermore, the surgical Short List was already in use for formal international harvesting of data.[7] The governing surgical societies have, therefore, not wished to compromise this to date by introducing new terms, which would not serve this particular purpose. Very little alteration of the surgical Short List was permitted. This has meant that the current listings are based upon the Short List of the Congenital Heart Surgery Nomenclature and Database Project as published in 2000,[2] with a few minor recent modifications.[7] It also includes some modifications which are, as yet, unpublished. The most significant of these are the inclusion of the first sub-hierarchy of ventricular septal defects, as detailed in the Long List of the *Annals of Thoracic Surgery* supplement,[8] and the dropping of the appendage *NOS* from all items. This latter reflects the opinion of the Executive of the Nomenclature Working Group that the generic term on its own was self explanatory, without the need for this or other clarifying nomenclature, such as *unspecified*, being affixed (see below).

In contrast, the Association for European Paediatric Cardiology had intended their Short List to be a comprehensive condensation of the Long List so that it would still be possible to code all lesions and interventions to a reasonable level of accuracy. It was also intended to be a solution to database objectives of a wide range of specialists within paediatric cardiology and cardiac surgery. Thus, it was logical to ensure that all items in the Short List of the Heart Surgery Nomenclature were catered for thoroughly in the European Paediatric Cardiac Code. The Coding Committee of the Association for European Paediatric Cardiology, with ratification by its Council, therefore agreed to incorporate, and map accurately, all of the terms in the Short List of the Congenital Heart Surgery Nomenclature and Database Project which were not represented in the European Short List. The European Paediatric Cardiac Code Short List has, therefore, been substantially modified. It now contains 317 additional items. The map to the Short List of the International Congenital Heart Surgery Nomenclature and Database Project system contains 51 entries where there was *no exact equivalent* term in the latter system.

In the crossmap of the Short List of the Congenital Heart Surgery Nomenclature and Database Project to that of the European Paediatric Cardiac Code, almost all of the entries in the surgical Short List have an equivalent term within the European Paediatric Cardiac Code, with the exception of those entries within the surgical Short List which contained the word *Other* (for example: *Coarctation repair, Other*). It was noted by the Executive of the Nomenclature Working Group that the use of the word "other" could confer different meanings to a term depending on the list in which it was included, that is the Short versus the Long List, and therefore these items should not be mapped. For instance, *Coarctation repair, Other* on the surgical Short List implies the use of a technique for surgical repair different from the types listed: end-to-end, end-to-end extended, subclavian flap, patch aortoplasty, and interposition graft. Within the surgical Long List,[9] the same entry *Coarctation repair, Other* is included in a list of routes by which the repair can be performed: left thoracotomy, right thoracotomy, median sternotomy, and using a transcatheter technique. The selection of *Coarctation repair, Other* from the Long list implies, therefore, a different meaning than the selection of the same term from the Short List. In addition, any entry containing the appended term *Other* may change meaning over time, as additional terms are added to the parent list from which the term was derived. The more comprehensive nature of the Short List of the European Code also meant that there was frequently more than one potentially equivalent term in this list for a corresponding item in the surgical Short List. For example, *Pulmonary valve, Other* in the surgical Short List could be mapped to any of *Pulmonary valvar abnormality* (09.05.00), *Pulmonary valvar abnormality – acquired* (10.35.01), *Post-procedural pulmonary valvar complication* (15.30.00) or *Pulmonary valvar prosthesis complication*

(15.30.08), with the inevitable potential for inaccuracy if trying to submit data from an institution using the surgical nomenclature to a database based upon the European Code. As a consequence of these factors, all terms from the Short List of the International Congenital Heart Surgery Nomenclature and Database Project with the addition *Other* have been entered as *no exact equivalent* in the crossmap of the item to the Short List of the European Paediatric Cardiac Code. It is anticipated, however, that in the near future there will be a series of further changes in the surgical Short List, which will address some of these issues. It is hoped that it will then be possible to produce a fuller and more accurate crossmap in this direction.

Crossmapping issues and the development of crossmapping rules

It can be seen from the above discussion that the crossmapping process has allowed further clarification of several issues concerning nomenclature and databases that have been difficult to resolve. Four of these issues that have been further clarified are discussed in this review:

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

Optimal performance from systems of nomenclature can be expected in an environment where the database, or system for entry of data, has certain standard regulations and requirements. The person entering the data, the nomenclature coder, must be forced to choose from the choices in the list of nomenclatures, and not be allowed to type free text directly into the fields for "Diagnoses" and "Procedures". A separate "Comments" field will then allow further free text to add additional description to any individual diagnosis or procedure that has been chosen. The crossmapping, and the systems themselves, will work effectively in environments that follow this basic rule or principle. This fundamental principle also leads to logical solutions for the first two issues above.

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

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

When discussing cardiac chambers, such as atriums and ventricles, and spatial relationships, the words *left* and *right* can be confusing. Rules were therefore created to provide consistency and accuracy of descriptive terms of anatomical phenotypes. For cardiac chambers, unless otherwise stated, *left* refers to morphologically left, and *right* refers to morphologically right. Thus, left ventricle means the morphologically left ventricle, left atrium refers to the morphologically left atrium, and right atrial appendage refers to the morphologically right atrial appendage, and so on. When discussing cardiac chambers, the words *left* and *right* do not imply sidedness or position. If one wishes to describe the position or sidedness of a cardiac chamber, it is necessary to use terms such as *left-sided ventricle*. The term left ventricle, therefore, merely means the morphologically left ventricle, and does not mean or imply left-sidedness or right-sidedness. Similarly, it does not imply connections to the right or left atrium, or the pulmonary or systemic circulations. In contrast, when describing the superior caval vein, and using the

prefix *left* or *right*, it is the spatial position that is being alluded to, rather than any other connection or phenotypic variation that may exist.

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

The future of the mapping process

The process of crossmapping the existing nomenclatures represents a work in evolution, which currently is encompassing the respective Long Lists of the two systems.[3] As discussed above, the process has allowed further clarification and understanding of several controversial issues. The process and our understanding of these issues will continue to evolve, as may the definitions, principles and rules. In turn, this will lead to further revisions in the lists, such as removing the terms *NOS, Other, (unspecified)* and *(DESCRIBE)* from the hierarchies, as discussed above. As the mapping process progressed, it became evident that, in general, it was easy to add new terms to the systems, but more challenging to merge or drop terms. A mechanism is evolving through the joint efforts of the Society of Thoracic Surgeons and the European Association of Cardiothoracic Surgery to facilitate these updates to the surgical lists. A similar mechanism is already in place within the Association for European Paediatric Cardiology for the European Paediatric Cardiac Code.

In fulfilling its first mandate, the Executive of The International Working Group for Mapping and coding of Nomenclatures for Paediatric and Congenital Heart Disease examined the problems, proposed solutions, and established the groundwork necessary for completing the second mandate of the working group. This was to crossmap the respective Long Lists of the European Paediatric Cardiac Code and the International Congenital Heart Surgery Nomenclature and Database Project, and/ or develop a single "super-tree" of nomenclature by the time of the next World Congress in Buenos Aires, Argentina in 2005, as outlined in the related article in this issue of *Cardiology in the Young*.[3] It is hoped that the ultimate outcome of this work will be a comprehensive list of entries for paediatric and congenital cardiac disease, as constructed with broad input from cardiac societies and associations from around the world. This will then be the International Paediatric Cardiac Code. It is anticipated that this will then become the universal standard coding system for databases worldwide, enabling truly representative and validated global multicentric research, which in turn, will lead to improvements in the care and treatment of patients with paediatric and congenital heart 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[2]) are available on the following sites, respectively:

www.aepc.org,
www.pediatric.ecsur.org, and
www.sts.org.

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