Special Report: Coding of Congenital Heart Defects in the EUROCAT Database

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The EUROCAT Coding and Classification Committee have over many years developed tools to improve the malformation coding within EUROCAT. These tools have to be continuously updated and improved following changes and progress in diagnostics and treatment. For the Joint Action contract 2011-2013 one of the tasks were:
Update EUROCAT CA subgroups with special emphasis on Congenital Heart Defects subgroups, update classifications and definitions of CA, disseminate coding guidelines and respond to coding queries.

Classification systems used for congenital heart defects:

There are several published classification systems for congenital heart defects (CHD).

The Baltimore-Washington study 1981-83 (1) used the classification system from International Society of Cardiology. All cases in the study were manually reviewed by a paediatric cardiologist.

In the review paper from Hoffmann in 2002 (2) CHD was defined as "A gross structural abnormality of the heart or intrathoracic great vessels that is actually or potentially of functional significance" using the old definition from Mitchell in 1971. CHD cases were grouped in three groups: severe, moderate and mild. ASD and VSD could be classified as either moderate or mild, which means that more than one malformation code is needed for classification.

In a study from Northern Region UK (3) CHD cases were grouped in three groups: complex, significant and minor. A two-dimensional classification system was used and required manual validation of the diagnosis for inclusion in the three groups.

The Atlanta Birth Defects Programme (4) received cases coded in ICD9 (six digits) with additional information on cardiac examinations, treatment and surgery. Paediatric cardiologists manually reviewed all cases and re-coded to Congenital Heart Surgery Database system (STS).

There are two coding systems developed by paediatric cardiologists: one from the Association for European Paediatric Cardiology (www.aepc.org) and one from an international nomenclature working group (www.ipccc.net). Both coding systems are much more detailed than ICD10 and require detailed information about echocardiography and other cardiac examinations. The Paris EUROCAT registry has evaluated the IPCCC coding in a publication (5). Eighty percent of the cases could be described with a single IPCCC code.

Conclusion: all of the above-mentioned CHD classification systems require much more detailed information on each case than available in EUROCAT.

Data for the survey on CHD coding in the EUROCAT Central database

The EUROCAT subgroups include 19 subgroups related to congenital heart defects: one including all CHD, 17 specified CHD subgroups and the severe CHD subgroup including 13 specified CHD subgroups.

Using this system, most ICD10 codes for CHD (Q20-Q26) are included in a specified CHD subgroup. The aim of this report is to describe the proportion of EUROCAT CHD cases that are not allocated to a specified CHD subgroup and give proposal for new CHD subgroups.

Data for this report is based on 2010 cases from 30 registries with data extracted from the Central database in autumn 2013. This means that most registries have updated the 2010 data with cases diagnosed in 2011. For small registries one year of data may not be representative for all years send to EUROCAT.

Total prevalence of CHD among the 30 registries ranged from 34.43 per 10.000 births to 140 per 10.000 with 5 registries above 100 per 10.000 births and 8 registries below 60 per 10.000 births (Figure 1). Prevalence of CHD cases not included in a specific CHD subgroup ranged from 0 to 19.71 per 10.000 births and from 0 % to 22 % of total cases (788 cases in total).

Description of cases not included in a specific CHD subgroup

A document including all CHD cases not included in a specified CHD subgroup has been prepared by Ruth Greenlees in September 2013, including 788 cases from 2010 and 429 cases from 2011. Number of cases not in a specified CHD subgroup from 2007 to 2009 ranged from 664 to 743 in the 30 registries.

The cases can be divided into 2 groups:
Other specified CHD (Q248) or unspecified CHD (Q249) - or another CHD code not in a EUROCAT CHD subgroup except all CHD subgroup

Number of cases coded with Q248 was 173 (22 %) and Q249 was used for 194 cases (25 %) of the 1217 cases in the list.

For other CHD codes the most important used codes were:
Q201double outlet right ventricle: 39 cases

Q202 double outlet left ventricle: no cases

Q205 corrected transposition: 9 cases

Q206 atrial isomerism – Ivemark: 5 cases

Q208 other anomalies of cardiac chambers and connections: 38 cases

Q209 unspec anomalies of cardiac chambers and connections: 20 cases

Q214, Q218 and Q219 other and unspec septal defects: 49 cases

Q223 other anomalies of pulmonary valves: 23 cases

Q232 and Q233 mitral valve anomalies: 84 cases

Q240 dextrocardia: 31 cases

Q245 anomaly of coronary vessels: 27 cases

Q250 PDA preterm: 70 cases (reported with other major anomalies)

Q252 interrupted aortic arch: 8 cases

Q253 supravalvular aortic stenosis: 29 cases

Q254 other anomalies of aorta: 126 cases

Q2541 persistent right aortic arch: 24 cases (add to list of minors?)

Q255 atresia of pulmonary artery: no cases

Q256 stenosis of pulmonary artery: 153 cases (add to list of minors?)

Q261 persistent left superior vena cava: 23 cases (add to list of minors?)

Q268 other anomalies of great veins: 23 cases

Conclusions and recommendations from Coding and Classification Committee November 2013

Half of all CHD cases not in a specific CHD subgroup are coded with Q248 and Q249. The value of a specific subgroup including these two codes will be very limited, as the CHD anomalies are either poorly specified or very rare. There may be some potential for data cleaning and improvements of coding among these cases.

The coding committee recommends adding three new CHD subgroups: double outlet right ventricle, mitral valve anomalies (Q232, Q233, only as primary anomaly), aortic atresia/interrupted aortic arch (Q252), all three to be included in severe CHD.

Further it is recommended to exclude Pulmonary artery stenosis (Q256) if GA <37 weeks and add this to the list of minors. Further to add persistent right aortic arch and persistent left superior vena cava to the list if minors in written text. When the subgroups will be revised, the codes for these two anomalies (Q2541, Q261) will be added to the list of minors. Some registries have a high overall CHD prevalence due to reporting of these anomalies as the only CHD.

These changes are expected to be implemented at the next revision of subgroups, time unknown.

A coding tip on PDA (include only if surgical/catheter treatment is performed/planned) will be written.

A coding tip on single ventricle has been written and further coding tips and validation routines in the EDMP will be written to improve the coding of complex CHD.

References

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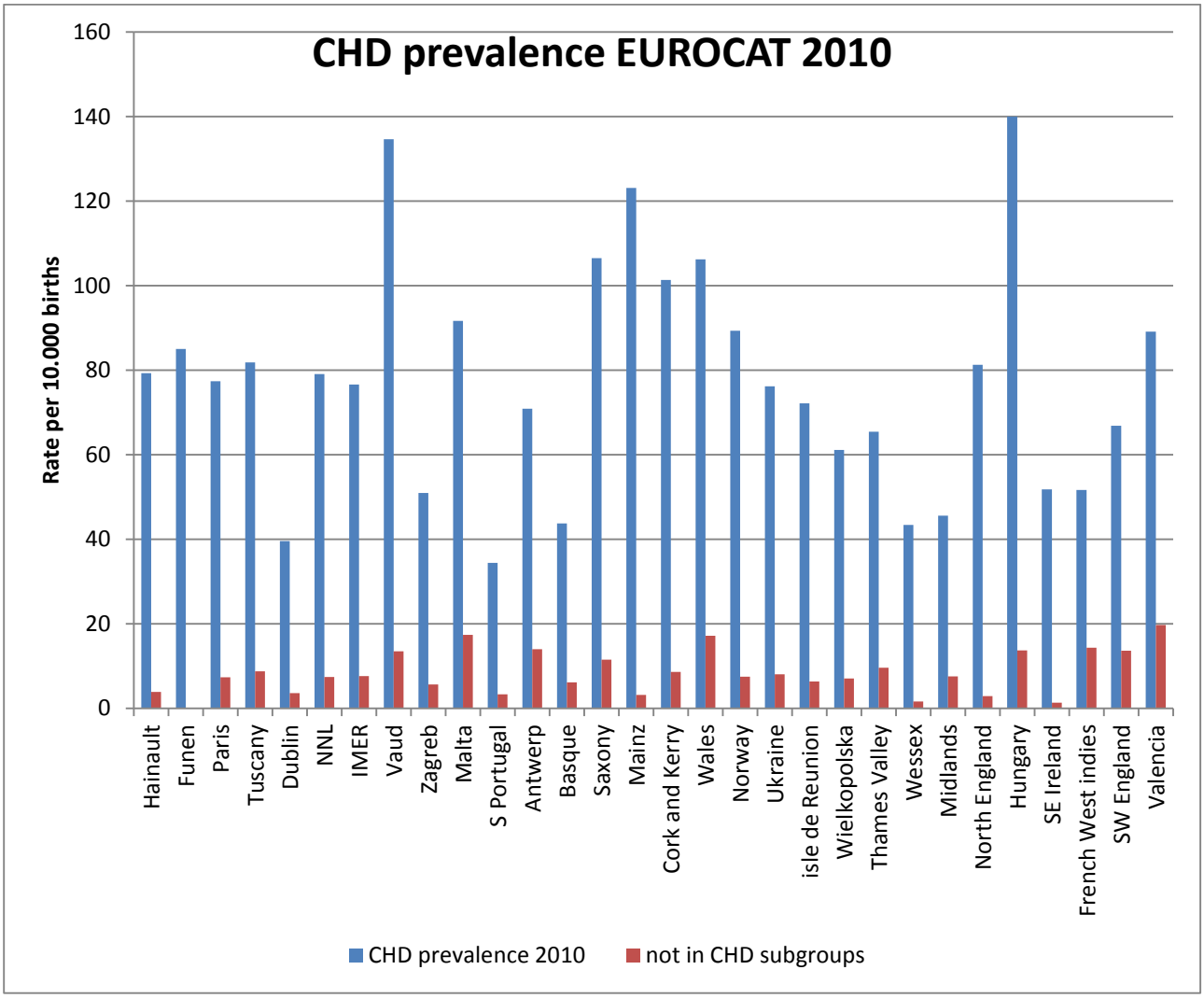
Figure 1