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Coding 299 Fetal Hearts Scans Using One Single Item from International Pediatric and Congenital Cardiac Code (IPCCC) and the Anatomic and Clinical Classification (ACC-CHD) Lists: Results, Limits, and Comparison of Discordances Related to Professional Experience

Abstract

Background: The international nomenclature of Congenital Heart Diseases (CHD) remains challenging. Classifications have been proposed such as the International Pediatric and Congenital Cardiac Code (IPCCC) and the Anatomic and Clinical Classification (ACC-CHD).

Objective: To Evaluate the clinical application of these two classifications on cardiac antenatal diagnosis.

Methods: We retrospectively included fetal echocardiograms over 6 years. Reports were independently coded with 1 single code by 3 pediatric cardiologists with increasing experience junior (J), senior I (SI) and II (SII). Discordances between doctors were compared to a gold standard code, with focus on coding difficulties and effects of professional experience using IPCCC and ACC-CHD.

Results: 180 scans were included. Using either "IPCCC short list 2012" or "ACC-CHD", coding with 1 item was difficult for SI and SII in 15% of cases. IPCCC was too exhaustive for its simple use leading to discordance. ACC-CHD was also difficult to use (learning curve, use of 1 code, complex CHD). Coding concordance using ACC-CHD main categories was higher for seniors compared to junior (J-SI, p= 0.04; J-SII, p=0.02), no differences between seniors. Compared to the gold standard for ACC-CHD (main, sub) categories, junior concordance was lower (73.3%, 71.1%) than SI (90%, 83.3%, p<0.005) and SII (88.3%, 87.2%, p<0.0001). Senior concordance was stronger (75%) with ACC-CHD sub categories compared to IPCCC (65%, p=0.028).

Conclusion: IPCCC and ACC-CHD remain difficult to use in clinical practice. Many functional abnormalities are not listed in the ACC-CHD but could be updated with a few more sub-groups to increase ease of use in this particular setting.

Keywords: congenital heart disease, nomenclature, antenatal diagnosis, classification

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Introduction

The nomenclature of congenital heart disease (CHD) is a real challenge, because of the variety of anatomies and increasing accuracy of descriptions of their structures.[1] The desire to classify and to code CHD has motivated the scientific community[2,3] for almost a century and is perhaps just as strong if not more so today.[4] Considering the number of adults with congenital heart disease (GUCH)[5,6] and the number of registries in pediatric and congenital cardiac diseases (EUROCAT,[7] EPICARD,[8]) standardizing CHD nomenclature is a scientific and clinical need. Since October 2000, the International Nomenclature Committee for Pediatric and Congenital Heart Disease brings together dozens of international expert centers for this purpose. This committee allowed the emergence of the International Pediatric Congenital Cardiac Code (IPCCC).[9] It offers a complete and comprehensive coding system (over 10,000 referenced codes). In 2011, a French team, in collaboration with other European centers, published a new CHD coding system, the Anatomical and Clinical Classification of Congenital Heart Defects (ACC-CHD).[10] Their goal was to achieve a more intuitive and easy to use tool, grouping heart diseases in 10 categories and 23 subcategories. Indeed, they might have been somewhat disconcerted by the heavy use of IPCCC, despite its unsatisfactory quality and accuracy.

Given the importance of the nomenclature in the analysis of congenital heart disease, it seems essential to use the tools at our disposal. However, today, it is unclear whether one coding system is more efficient than another. To our knowledge, no medical team has published a critical study evaluating these tools. The aim of our study was to compare the IPCCC and ACC-CHD systems in terms of their usefulness and reproducibility on a series of diagnoses of consecutive fetal heart diseases.

Methods

We performed a retrospective, single-center study, over the period from September 2006 to July 2013. Data collection was

centralized at the University Hospital of Martinique (Fort-de-France, French West Indies). All antenatal diagnoses were made by a single practitioner, specialized in antenatal echocardiography. These fetal echocardiographies were performed after a primary screening during conventional French monitoring of pregnancies (ultrasound at 10weeks of gestation (WG), 20WG and 30 WG) by competent midwives or by gynecologists. We decide to compare two coding systems, "IPCCC short list 2012" (EPCC short list) and "ACC-CHD", on a series of antenatal diagnoses. The authors of these encodings had not yet ruled on their clinical use in the field of fetal cardiology. Therefore, we applied their tools to the fetal series as if it was postnatal diagnosis.

Once all reported diagnoses had been collected and personal details removed (blind evaluation, without any information about the post-natal history), we presented them to three expert operators in congenital cardiology. One of the operators was a junior (J), the other two were senior doctors with experience ranging from 4 (SI) to 15years (SII). Each cardiac disease had to be coded with IPCCC and ACC-CHD using one and only one single code to facilitate comparison by the participating operators. This double blind coding (i.e. participants were not aware of the codes selected by the others, was performed according to the "good sense" of the encoder to determine the most prominent malformation. In any case, only diagnoses cod able in IPCCC and ACC-CHD were considered.

Finally two other independent senior practitioners, specialized in congenital heart diseases, determined the most appropriate encoding for each diagnosis (called Gold Standard / GS). For this, they used the definitions of heart disease published by the International Nomenclature Committee for Pediatric and Congenital Heart Disease (Definitions Working Group).[11] The 3 sets of codes, the two systems used (IPCCC and ACC-CHD), were compared with each other and with the gold standard. The differences between these encodings (inter-operator discordances and operators -gold standard discordances) were analyzed with Chi-2 test. Analysis was conducted using *Medcalc* (MedCalc Software, Mariakerke, Belgium).

Results

299 foetal echocardiographs were carried out during this period. Except for the years 2006 and 2013 (not complete) the standard deviation of the activity was 33-56 annual diagnoses. 187 foetuses had a foetal echocardiogram over this period: 32 foetuses had 2, 6 foetuses had 3, 5 foetuses had 4 and 2 foetuses had 5 ultrasounds. We considered that each ultrasound should be coded independently, because the diagnosis could have been modified from one ultrasound to another. The average term of diagnosis was 26.3weeks of gestation. There were 7 twins and 225 single pregnancies.

The three operators were able to encode 100% of diagnoses with IPCCC. On the other hand, using ACC-CHD items only 60.2% (180) of diagnoses could be coded by all three coders. As the main coding system ACC-CHD is based on 10 major categories (Table 1, taken from the bibliographic reference number 2), we were able to carry out a study of matches per category. In conclusion, 180 diagnoses could be coded in the two systems described, as opposed to 119 where this was not possible (Table 2). Characteristics of the population coded in both systems (180 diagnoses) are summarized in Table 3.

Comparisons between the IPCC and ACC-CHD codes assigned by the three different operators the IPCCC and ACC-CHD are presented in Table 4. Given the range of codes available for IPCCC, it was difficult to draw conclusions about the types of differences emerging between operators. However, using IPCCC, inter-operator concordance is quite similar between the three operators, around 65%. Concerning ACC-CHD, inter-operator matches were generally better than those of the IPCCC despite a statistically significant difference between operators, and a stronger correlation between more experienced operators (J versus SI-SII, p=0.03). Restricting the analysis to ACC-CHD categories only, the difference remains significant (p=0.04; J-SI = 73%, J-SII = 72%, SI-SII = 82%).

Operators-gold standard (GS) concordance for IPCCC and ACC-CHD are presented in Table 5. Again, there is a better match for the ACC-CHD. The operators-GS concordance for both coding systems is also favoured by experience (p<0.05). Again, looking to ACC - CHD families only compared to GS, we found the same statistical trend as for inter- operators comparison (p=0.02; J-GS = 73%, SI-GS = 90%, SII-GS = 89%).

Table 1 ACC-CHD classification with 10 main groups

Group
1. Heterotaxy, including isomerism and mirror-imagery
2. Anomalies of venous return
3. Anomalies of the atria and interatrial communications
4. Anomalies of the atrioventricular junctions and valves
5. Complex anomalies of atrioventricular connections
6. Functionally univentricular hearts
7. Ventricular septal defects (VSD)
8. Anomalies of the ventricular outflow tracts (ventricular-arterial connections)
9. Anomalies of the extrapericardial arterial trunks
10. Congenital anomalies of the coronary arteries

Table 2 Impossibility of use ACC-CHD (119 cases)

Diagnostic Doubts	Ventricular Asymmetry	Unclasssified (Cardiomegaly, Tumor, Pericardial Effusion,…)	Normal	Extra-Cardiac	Heart Rhythm Abnormalities	Total
7	15	20	52	3	22	119

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Table 3 Epidemiological characteristics of the population (180 scans)

	Total of 180 Scans
Number of patients	134
average age of mothers (years)	30.1
Mean weeks of gestation (WG) at the time of ultrasound	26.3
Singletons / Twin	177 / 3
Medical termination of pregnancy	34
Fetal Death	2
Stillborn	2
Newborn living at birth	96

Table 4 Inter-operator concordances

	ACC-CHD Concordance			IPCCC Concordance		
Population	J-SI	J-SII	SI-SII	J-SI2	J-SII2	SI-SII2
Number	127	126	139	121	116	117
Percentage	70,56	70	77,22*	67,22	64,44	65

*p=0, 03 (Chi-2), comparing SI-SII concordance with the two others concordances (J-SI et J-SII) for ACC-CHD.

Table 5 Operators-Gold Standard Concordances

	ACC-CHD Concordance			IPCCC Concordance		
Population	J-GOLD	SI-GOLD	SII-GOLD	J-GOLD	SI-GOLD	SII-GOLD
Number	129	154	158	112	128	137
Percentage	71,67*	85,56	87,78	62,22*	71,11	76,11

*p<0, 05 (Chi-2), comparing J-GOLD concordance with the two others concordances (SI-GOLD et SII-GOLD) for ACC-CHD and IPCCC.

Furthermore, closer analysis of the coding results under ACC-CHD, revealed that some categories (Figure 1), such as category 6 (Functionally univentricular heart) and category 9 (Anomalies of extrapericardial arteries trunks) the highest rate of discordance. There are no errors listed for category 3 (anomalies of the atria and interatrial communications), 5 (complex abnormalities of atrioventricular connections) and 10 (congenital anomalies of the coronary arteries) because using a single main code, our series had no diagnosis for these categories.

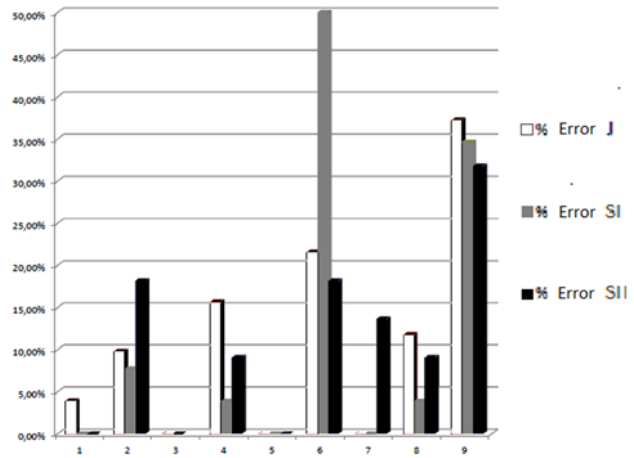

Figure 1 The three encoders are represented: the junior (J), the senior I (SI), the senior II (SII).

Discussion

We evaluated the applicability and reproducibility of two coding systems (IPCCC and ACC-CHD) for fetal heart diseases, in a single inter regional center. Regarding the applicability, 100% of diagnoses have been coded by IPCCC compared to 60.2% by ACC-CHD. The absence of ACC-CHD code for foetal rhythmic disorder, conduction disorders, tumoral pathologies, pericardial disease, or even just a normal cardiac structure, greatly limit the extension of this particular coding system to all the pathologies observed in foetal cardiology (see Table 2), in comparison with the IPCCC. Regarding reproducibility, we observed a strong disparity between operators whatever the coding system used. However, the ACC-CHD was more concordant than IPCCC ($p<0.05$), for all types of comparison (inter-operator, operators-GS, family ACC-CHD). Nevertheless, we got the feeling that the ACC-CHD could represent a more "practical" or intuitive coding system than IPCCC. Looking at error analysis, several features have had a net impact, such as:

a. The type of coding system. Indeed, the ACC-CHD seemed more reproducible than the IPCCC, although we were not able to code more than 60.2% cases using this coding system.

b. The experience of the encoder. The junior operator had a greater rate of discordance with the two seniors ($p=0.03$) and with the gold standard ($p<0.05$).

In addition, we believe that the additional remarks about these coding systems may be helpful in stimulating discussion in the medical community:

I. First of all, there is clearly a need for a more unified and reproducible coding system. Neither the ACC-CHD nor the IPCCC systems seems optimal for clinical practice. A computerized segmental analysis might meet the demand for a coding system in congenital and paediatric cardiology which would be simultaneously easy to use, accurate and reproducible.

II. The need for a common international definition of CHD. In order to optimize the usefulness of these coding systems, it is important to remember their fundamental characteristics. A Coding system per se is objective but its interpretation for implementation remains subjective. Implementing the use of IPCCC without first reading the work of the Definitions Working Group[1] about (for example) hypoplastic left heart syndrome,[12] double outlet right ventricle,[13] or Ebstein's anomaly,[14] would be like "using a code

without reading the manual". This allows for a "common core" to define heart defects, but coding in clinical practice requires heavy and demanding work for medical teams wanting to "speak the same language."

III. The need for a reliable ultrasound evaluation thanks to which the operator would be free of "doubts". In order to implement a coding system, clinicians establishing a diagnosis should endeavour to minimise inaccuracies or the expression of doubt in their conclusions; however, this is a difficult challenge in daily clinical practice in such a difficult field as foetal heart scan. It would nevertheless be critical in allowing the most accurate and complete diagnosis possible by foetal cardiology teams using the IPCCC or ACC CHD coding systems.

IV. Coding overlaps in IPCCC. Regarding IPCCC coding in particular, it would have been logical to find complete agreement between operators. This coding system is fully complete and accurate, and the variability should have been zero. However, we found that the complicated nature of the system and multiple over lapsled to differences in choice of coding. For example, 3 codes might describe a "normal" heart: "normal heart" [01.01.00] "absence of fetal heart abnormality detected" [14.10.00] "Foetal echocardiography study: no abnormality detected " [12.14.00]).

V. Applicability to the foetus? The applicability of these coding systems to antenatal series remains to be discussed. The authors of these systems never exclude formally antenatal diagnosis of their application. However, we found that some recurring prenatal diagnoses were suffering from a lack of information (or presence) in these coding systems. As already explained, the normal heart is not available in ACC-CHD while different codes exist in IPCCC. As such, we specify that codes do exist in IPCCC for the foetal period but they are more succinct. Using ACC-CHD, ventricular imbalance was often coded in a first instance by the junior operator 6.3 (Ventricular imbalance with dominant hypoplastic RV and LV) but almost never encoded by Senior operators who considered that category 6 (Functionally univentricular hearts) should not contain the term ventricular imbalance unless one ventricle is already clearly hypoplastic. In contrast to IPCCC considerations, we regret that the authors of the ACC-CHD were unable to better define the terms used in their coding.

Finally, we found that coders can have different types of approaches in applying these coding systems. Some will have a functional logic (for a therapeutic approach) others will have anatomical, embryological logic, or even physiological sense... This is why the authors of the ACC-CHD recommend to use their tools with "logic and intuition"(10), or that the authors of the IPCCC proposed thousands of codes to be combined with each other. We feel that the clinical truth probably lies somewhere in between. Our study noted some strengths and limitations for each coding system, while finding a relatively disappointing inter-operator concordance. Better concordance is likely to come from improving these coding systems and the learning curve for teams using them. Ultimately, it seems essential that teams wanting to apply these coding systems agree on a standard system to "choose" the most appropriate code for each heart disease.

The limitations of our study are: 1) we only used one single code for IPCCC and ACC-CHD, to simplify the reproducibility study, but neither coding system has been clearly designed to do so; 2) Only one operator performed all diagnoses, using sometimes inaccurate or subjective terms (Table 3); 3) both senior operators encoded in a "clinical / therapeutic" approach, while the junior operator encoded in an "anatomical" approach; 4) the three operators had studied the tools in varying ways (full articles, definitions of the Working Group) before using them 5) our study included prenatal data, without being sure that these encodings were fully adapted for this type of diagnosis.

Conclusion

We believe that working to improve these coding systems is a necessity and a priority for congenital and paediatric cardiology. Getting better tools to compare worldwide activity, studying groups of patients, providing reliable and robust statistics to patients and relatives, should motivate us to continue in this direction. We hope that other clinical teams, like ours, will try to apply and critique these tools to refine and propose adjusted versions.

Authors contributions

I. Olivier Villemain: Concept/Design, Data collection, Data analysis/interpretation, drafting article, Critical revision of article, Approval of article.

II. Alexandre Bretonneau: Data collection, Statistics, Concept/Design, Data analysis/interpretation, Critical revision of article, Approval of article.

III. Michelle Gueneret: Data collection.

IV. Laurence Long: Data collection.

V. Jean-Marc Rosenthal: Data collection.

VI. Damien Bonnet: Critical revision of article, Approval of article.

VII. Hugues Lucron: Concept/Design, Data collection, Data analysis/interpretation, Critical revision of article, Approval of article.

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Conflicts of interest

Author declares there are no conflicts of interest.

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