Educational Review

Diagnostic Nomenclature and Expression Formats for Cardiac Malformations: Confusions, Controversies, Conflicts, Convictions, Whatsoever!

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Cardiac malformations are described using several formats of expression, mainly based on three different backgrounds; that is, either from the aspect of embryological development, via a morphological approach, or by means of clinical classifications. We come across occasionally, or probably better to say often, a circumstance in which people misunderstand or are confused with diagnoses of the heart lesions. Common nomenclature for congenital heart diseases is nearly achieved, but not complete yet. This on-going attempt is also being accompanied by the developing coding system. The coding system might not cover all information the heart team needs to share in really practical settings. As long as the way how to describe heart lesions is not perfectly unified, we have to communicate reasonably well realizing that there are several standpoints for diagnoses of cardiac malformations. Behind each word, there are underlying thoughts we should note. Words are derived from concepts, and, at the same time, promote the concepts.

Keywords: cardiac morphology, diagnosis, heart malformation, sequential segmental analysis, congenital heart disease

Morphology and Embryology in Congenital Heart Disease —Don't You Have a Friend Like This?

One day, George sat in a lecture room together with his peers, awaiting the arrival of a professor of cardiac morphology. The professor was famous for a systematic approach to analyzing the structure of heart defects. George had only a limited knowledge of congenital cardiac malformations. He attempted to read a textbook on cardiac embryology three years ago when he was a medical student. He remembered that he was not able to complete any of the chapters. Each time he started from the beginning of a chapter to make sure that he could understand perfectly, the book always turned out to be a nice pillow on his desk within 20 minutes. To him, pictures were just like abstract artistic shapes, their titles being labelled in Latin. On the basis of this discouraging memory, he was hoping that today's lecture would not be a further setback.

"Good morning, ladies and gentlemen." The professor began his talk brightly. With his loud and clear voice, I could not sleep through his lecture, George thought. The professor explained very precisely the architecture of the normal heart. Not too fast, not too detailed, not too specialized. His interactive presentation created a nice atmosphere. Subsequently, he moved on to one example. He showed how to analyze the abnormally structured heart step by step. At the end, the descriptive list on the specimen became extensive (Table 1a). George realized that he needs some verbal tools to find out features of a structure in detail. To recognize an object is rather different from just to see it; the former needs active participation of mind, he thought. When George went back home, however, he was not entirely sure whether he could recall all the key points the professor demonstrated during the day and whether he could apply the principle to each case he might have at the time of exam-

J-STAGE Advance Published Online: January 31, 2024

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doi: 10.24509/jpccs.23-006

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Table 1 An example of diagnostic descriptions on the basis of three major formats

(a) Purely morphologic basis	(b) with embryologic background	(c) for hasty clinicians
usual atrial arrangement - morphologically right atrial appendage on the right and left atrial appendage on the left - right superior caval vein connected to the right atrium	situs solitus - SVC and IVC to right atrium on the right PVs to left atrium on the left	
- left superior caval vein returning via the coronary sinus - inferior caval vein connected to the right atrium - pulmonary veins connected normally to the left atrium	- left SVC to right atrium via coronary sinus	- PLSVC
- small atrial septal defect at the oval fossa	- small ostium secondum atrial septal defect	- restrictive ASD
univentricular atrioventricular connection - absent right atrioventricular connection - the left atrium connected to the dominant morphologically left ventricle	tricuspid atresia [SDD]	TA/TGA (type IIc)
- cleft in anterior leaflet of left atrioventricular valve - the small and incomplete morphologically right ventricle on the right anterior to the left ventricle - right hand ventricular topology	- cleft mitral valve - d-loop ventricles	- mitral cleft
- small interventricular communication - ventricular apex pointing to the left	- small bulbo-ventricular foramen - levocardia	- restrictive VSD
discordant ventriculo-arterial connection - the aorta arising from the right ventricle and the pulmonary trunk arising from the left ventricle	transposition of great arteries	
- the aorta right anterior to the pulmonary trunk - pulmonary trunk larger than ascending aorta	- d-loop great arteries	- d-TGA
- narrow subaortic channel	- subaortic stenosis	- SAS
coarctation of the aorta - three neck vessels from the aortic arch	pre-ductal coarctation	CoA
- narrow patent arterial duct	- narrow patent ductus arteriosus	- small PDA
- aortic arch left to the trachea - normally branching pulmonary arteries	- left aortic arch	- left arch

An example of the so-called classical "tricuspid atresia" is documented. A morphological stance provides a precise list of sequential segmental analysis, but extensive and somehow verbose. An embryological approach demands certain knowledge in this field, and it may not be straightforward to understand the entire view. A clinical shorthand sounds concise and fashionable; still its users should pay attention to historical evolution of background ideas or the potential of misunderstanding/miscommunication.

ination. Therefore, he decided to buy a book written by the professor. He actually read the book with great interest. The way of thinking termed "sequential segmental analysis" seemed to be a good conceptual tool to him. He felt that everything would become clear-cut when he used such a systematic and scientific approach.

One year later, George had an opportunity to attend a short course on congenital heart disease abroad. He was hoping to augment his knowledge in addition to the morphologic approach he had learned. He did not suspect that topics would be discussed without using the "sequential segmental analysis" taught by the professor. Therefore, he was quite shocked to find that he could not understand completely what people were talking about. "Bilateral conus," "endocardial cushion," "Malposition of the great arteries," "SDL," "polysplenia," and so on. He felt he was on another planet. He wondered if everything might have changed for diagnoses of cardiac malformations during the past 12 months. After finishing the afternoon session of the first day, he asked one of the young participants in a friendly manner. "I did not understand very well the terminology the lecturers used today. Where were they from? What does l-loop mean?" "Well, you need to study more about the background of heart defects. Congenital malformations cannot be understood without knowledge of embryology, you see," the young man replied bluntly. "Embryology!" George exclaimed in his mind. "That's what I could not cope with in the past," a feeling of despair took the shine out of his eyes. After supper, he was still reflecting on what he had listened to during the day. "Am I hopeless? Is this field beyond my ability? But, probably, I could have understood some of the contents with knowledge of "sequential segmental analysis," couldn't I?" That night, George had a nightmare in which he was confronted with a menu written in Arabic characters and unable to

order anything for dinner.

Next day, the morning session was all right; imaging modalities were explained on relatively simple diseases. At lunch time, a senior lecturer sat next to George and gently asked, "How is the course going? Everything clear?" George replied frankly, "I was confused yesterday. I do not have enough knowledge of embryology. I could not understand the terminology and underlying concepts very well. I previously learned the basis of sequential segmental analysis. Is that completely useless?" The gentleman smiled and quietly said, "Embryology is an important aspect in order to understand congenital diseases. Providing background knowledge allows us to interpret and explain features of the malformed hearts. Having said that, it is not the only approach we have. Purely morphological description can, in practice, show us detailed architecture, whereas other designated stereotypes are also clinically relevant. I have to admit there are conflicts within the nomenclatures because we intend to establish academic terms as precisely and as specifically as possible. Still, the essence of these scientific dogmas exists commonly in the ultimate understanding of congenital heart diseases. By the way, did you know that the systematic approach to cardiac segments and their sequence was derived from an embryologic context initially?" "Interesting," said George. "In other words, the same condition is described in different languages according to academic standpoints, isn't it?" "That's correct." "Diagnoses made on a common matrix would have been easier for a beginner like me to understand. How do you describe this sort of malformation for example?" George illustrated an example that was nicely explained before on the basis of morphology. The gentleman explained his thought and its background patiently (Table 1b). George thanked him greatly for the fruitful personal tuition.

Soon after starting specialist training in cardiology a couple of years later, George came across a patient with malformations similar to that memorable case. When George began to present the morphologic diagnoses to a surgeon, he impatiently cut George's words short. "TA/TGA and CoA, isn't it? (Table 1c). How restrictive is the VSD? Is the aortic valve bicuspid? The aortic arch hypoplastic? Body weight? On prostaglandin? Lactate level? ……" George was flustered. The surgeon immediately taught George what to say, how, and why. "The expression you attempted initially should be precise, but it is far too long-winded for a busy surgeon like me," he added impishly. "If you presented the case at a scientific meeting in your manner, you would use half of your limited presentation time just for the morphologic diagnoses!" This time, George was not frustrated. "Another stance," he muttered to himself. Beyond accuracy, well-summarized diagnosis and clinical key points were needed. He also learned that attention should be paid to presence or absence of potentially possible impediments.

George has expanded his knowledge on many fronts. He experienced cardiology, particularly for congenital heart disease, at several institutions all over the world. He came to realize that, not only nomenclature of diagnoses, but also background concepts and practical skills of treatments were really varied. He felt people were occasionally on quite different wavelengths. "Sometime, we might be able to build up a consensus in a true sense. At present, at least, we need to prepare and to widen the scope for deeper knowledge so as to translate each opinion into a meaningful and non-misleading one," he calmly said to his new trainee.

Terminology for Congenital Heart Disease —Do You Need a Dictionary?

It should be useful to have a definitive dictionary of terminology in congenital heart diseases. As is often the case with dictionaries, for example between two languages, different grammars and background cultures make it very difficult to give exacting one-to-one correspondence in everything. No dictionary guarantees perfect translation on its own. Nonetheless, some examples are provided by listing similar, or comparable, expressions in comparison (Table 2).

Clinical Classifications of Congenital Heart Disease—Can You Memorize All Alpha-Numeric Stereotypes?

To classify stereotypes or subtypes of a spectrum of malformations, headings were given consisting of abbreviations and figures in the past (Table 3). Some people continue to use such headings with a sense of pride that they have studied meticulously the past literature, while others avoid using insipid typification with the explanation that they are confused with the digits and alphabets. If a classification is not well adopted, communication and understanding of real malformations are difficult.

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Table 2 Different terminology

Morphological stance	Embryological stance
Based on the nature of the appendage	Based on veno-atrial connection and splenic status
usual atrial arrangement	situs solitus
mirror image arrangement	situs inversus
right isomerism of atrial appendage	asplenia
left isomerism of atrial appendage	polysplenia
	(situs ambiguous no longer used)
[each of four headings not always corresponding to that of the other group]	
biventricular atrioventricular connection	
concordant atrioventricular connection	[S, D, ( )] or [I, L, ( )]
discordant atrioventricular connection	[S, L, ( )] or [I, D, ( )]
exceptional patterns in isomerism	
univentricular atrioventricular connection	
absent right atrioventricular connection	tricuspid atresia
absent left atrioventricular connection	mitral atresia
(no connection either to morphologically left or right ventricle)	(irrespective of right or left, depending of ventricular looping)
double inlet left ventricle (small and incomplete right ventricle)	single LV
double inlet right ventricle (rudimentary left ventricle)	single RV
double inlet indeterminate ventricle (true single ventricle)	(double inlet for separate valves, common inlet for a common valve)
(either through separate valves or a common valve)	common ventricle (very large VSD)
concordant ventriculo-arterial connection	[S, ( ), N]
discordant ventriculo-arterial connection	transposition of great arteries, d-TGA, l-TGA
double outlet right ventricle (50% rule)	malposition of great arteries, d-MGA, l-MGA
double outlet left ventricle (50% rule)	cono-truncal criss-cross
single outlet with pulmonary atresia	tetralogy of Fallot with pulmonary atresia
(aorta either from left or right ventricle)	DORV/TGA with pulmonary atresia
single outlet with aortic atresia	aortic atresia
(pulmonary trunk either from left or right ventricle)	
common arterial trunk	persistent truncus arteriosus
atrioventricular septal defect (AVSD)	common atrioventricular canal
(common atrioventricular valve)	endocardial cushion defect
(separate atrioventricular valve)	(complete/partial/intermediate/variant form)
(ASD component present, no VSD component)	ostium primum defect
complete transposition	
usual/mirror imaged atrial arrangement with concordant atrioventricular and discordant ventriculo-arterial connections	d-transposition, TGA [SDD]/[ILL], simple TGA
congenitally corrected transposition	
usual/mirror imaged atrial arrangement with discordant atrioventricular and ventriculo-arterial connections	l-transposition, TGA [SLL]/[IDD]
others	
usual/mirror imaged atrial arrangement with concordant atrioventricular and ventriculo-arterial connections with abnormal aorto-pulmonary orientation	anatomically corrected malposition (ACM)
usual/mirror imaged atrial arrangement with concordant atrioventricular and discordant ventriculo-arterial connections with aorta and pulmonary trunk normally oriented	posterior TGA
double outlet right ventricle (DORV)	
one of the four patterns in ventriculo-arterial connection (50% overriding rule: defined as RV giving rise to more than 50% of each of the semilunar valves.)	caused by incomplete absorption of subaortic conus
variations in relation to VSD orientation	variation in relation to aorto-pulmonary orientation
subaortic VSD	[SDD]/[SDL]
subpulmonary VSD	true or false Taussig-Bing, complex TGA
doubly committed VSD (outlet septum lacking)	
non-committed VSD (remote from either aortic or pulmonary)	
tetralogy of Fallot with DORV can coexist	DORV cannot coexist with tetralogy of Fallot
ventricular septal defect (VSD)	
juxta-arterial and doubly-committed VSD	distal conus defect
	supra-cristal VSD
atrial septal defect (ASD)	
oval fossa type atrial septal defect	ostium secondum ASD
sinus venosus interatrial communication	sinus venosus ASD
ventricular structures	
outlet septum	conus/infundibular septum
septo-marginal trabeculation	trabeculo-septo-marginalis
interventricular communication/foramen	bulbo-ventricular foramen
ventriculo-infundibular fold	subaortic conus

A morphological stance and an embryological stance do share many terms to describe heart malformations in detail. Some expressions, nonetheless, are rather discordant with each other. The different wording reflects different processes and backgrounds of thoughts.

Table 3 Clinical shorthand

Abbreviations and categories	Stereotypes
TA[1]	tricuspid atresia
I	concordant ventriculo-arterial connection (Aorta from LV)
II	transposition of great arteries (Aorta from RV)
-a	with pulmonary atresia
-b	with pulmonary stenosis
-c	no pulmonary stenosis
III	absent left atrioventricular connection (RA opening to LV)
TGA[2]	transposition of great arteries
I	complete transposition with intact ventricular septum
II	complete transposition with ventricular septal defect
III	transposition with ventricular septal defect and pulmonary stenosis (overriding aorta)
IV	transposition with intact ventricular septum and pulmonary stenosis (non-dynamic obstruction)
Patterns of the coronary arteries	based on
Shaher[3]	investigation using morphologic specimens
Yacoub[4]	surgical experiences
Leiden convention[5]	investigation using morphologic specimens
Boston group[6]	surgical series
PTA[7]	persistent truncus arteriosus
I	pulmonary arteries originating via a common channel from truncus
II	pulmonary arteries close together originating directly from truncus
III	pulmonary arteries originating bilaterally from truncus
IV	pseudo-truncus (nowadays understood as tetralogy of Fallot with pulmonary atresia and major aorto-pulmonary collateral arteries (MAPCA)
A1-4, B (Van Praagh)[8]	depending on VSD, pattern of PA branching, and interruption of the aortic arch
IAA[9]	interruption of aortic arch
A	all three neck vessels from the aortic arch
B	interrupted between left carotid artery and left subclavian artery
C	only brachiocephalic artery originating from the ascending aorta
CoA[10]	coarctation of the aorta
post-ductal, pre-ductal 1, 2, 3 (Keith)	depending on location and hypoplasia of the aortic arch
TAPVC[11]	totally anomalous pulmonary venous connection
I	supra-cardiac type (draining via the ascending vertical vein)
a	all four pulmonary veins to the brachiocephalic vein
b	all four pulmonary veins to the superior caval vein
II	cardiac type (draining to RA)
a	all four pulmonary veins to the coronary sinus
b	all four pulmonary veins directly to RA
III	infra-cardiac type (all four pulmonary veins draining via the descending vertical vein)
IV	mixed type (four pulmonary veins not forming a confluence)
cor triatriatum[12]	divided atrial chamber
I	accessory atrial chamber receiving all pulmonary veins and communicating with left atrium
A	no other connections (classical cor triatriatum)
B	other anomalous connections
1	to right atrium directly
2	with total anomalous pulmonary venous connection
II	accessory atrial chamber receiving all pulmonary veins and not communicating with left atrium
A	anomalous connection to right atrium directly
B	with total anomalous pulmonary venous connection
III	subtotal cor triatriatum
A	accessory atrial chamber receiving part of pulmonary veins and connecting to left atrium
1	remaining pulmonary veins connecting normally
2	remaining pulmonary veins connected anomalously
B	accessory atrial chamber receiving part of pulmonary veins and connecting to right atrium
1	remaining pulmonary veins connecting normally
single ventricle[13]	
A	LV type
B	RV type
C	common ventricle (large VSD)
tetralogy of Fallot	
I, II, III, IV, V (Kirklin)[14]	depending on features across the right ventricular outflow tract
I, II, III, IV, V (Kawashima)[15]	VSD location and manner of pulmonary stenosis/atresia
VSD[16]	ventricular septal defect
Kirklin I	juxta-arterial and doubly-committed (distal conus) type
II	perimembranous outlet/trabecular type
III	perimembranous inlet type
IV	muscular type

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Table 3 Continued

Abbreviations and categories	Stereotypes
ASD[17]	atrial septal defect
I	ostium primum type (atrioventricular septal defect, partial form endocardial cushion defect)
II	ostium secondum type
AP window[18]	aortopulmonary window
I	proximal type
II	distal type
III	total defect type
Valsalva sinus rupture[19]	
I, II, IIIv, IIIa, IV (Sakakibara-Konno)	depending on location of aneurysm and dommunication
Rastelli classification	in common atrioventricular valve
A, B, C[20]	based on patterns of leaflets and tension apparatus
LV-RA communication[21]	left ventricular-right atrial shunts
I	
II (A1, 2, 3, B1, 2, 3, 4)	
III	based on radiologic findings
Vascular ring[22]	branching patterns of the thoracic aortic pathway
I A, B	
II A, B, C	
III A, B, C	
IV	based on embryologic development

Clinical classifications of congenital heart disease have been proposed aiming to diagnose the lesions better and to treat the patients optimally. When using handy abbreviations and numerical/alphabetic typification, exact definitions should be understood behind those symbols. Otherwise, they are misleading.

Describing Congenital Heart Disease —Which Would You Like?

Any descriptions in the literature should aim to be as accurate, as suggestive, or as practical as possible. At the same time, each one has its downside.

Detailed morphological diagnoses are occasionally extremely long. Of course, to be precise is of utmost importance. But, it is a reality that some people feel reluctant to describe every aspect in long-hand, especially in this era of text-messaging. This is more often the case in those whose first language is not English.

On the other hand, embryology requires considerable effort for understanding. We need to accumulate bulky knowledge. Precise details of normal development are still being elucidated, let alone controversies in abnormal development. Obviously, brief embryologic terms imply background insight into development of the heart, some of which may be presumptive. Because of various levels of knowledge, communication between discussants and the audience is not always smooth.

Truncated expressions also need some background knowledge of the literature, and, more importantly, they are occasionally incomplete, confusing, or misleading. Such shorthand could be used within a clinical team, but not recommended in academic circumstances.

As long as no unified way is established, at least at present, we have to communicate reasonably well realizing that there are several standpoints for diagnoses of cardiac malformations. Common nomenclature for congenital heart diseases is being negotiated[23–26] or nearly established even, but not completely yet. This on-going attempt is also being accompanied by the developing coding system.[27–30] Behind each word, there are underlying thoughts we should note. Words are derived from concepts, and, at the same time, promote the concepts.

Conflict of Interest

The author has nothing to declare.

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