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How to describe complex congenital heart defects? A hierarchical approach for diagnosis, using the Anatomic and Clinical Classification of Congenital Heart Defects.

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Background: To describe complex congenital heart defects (CHD) in their diversity requires a common language, in order to unify the diagnostic process. As a first step, we designed the Anatomic and Clinical Classification of CHD (ACC-CHD) based on a multi-dimensional approach encompassing anatomic, diagnostic and therapeutic criteria. The second step aims to establish a hierarchy among the different components of a complex CHD, which is fundamental since it places the main lesion into one, and only one, of the 10 groups of the ACC-CHD.
Methods: The hierarchy within the 10 groups (G) and 23 subgroups of the ACC-CHD was based on the clinical aggregation process reflecting the routine practice of paediatric cardiology, but also on anatomy and criteria used for medical and surgical management. For example, transposition of the great arteries is the main defect (D1) when associated with a VSD but not when associated with double-inlet ventricle, because of the different surgical management: arterial switch versus univentricular approach. We applied this method to data acquired from a population-based cohort of patients with CHD in France (the Epicard study) made up of 2867 cases (82% live births, 1.8% stillbirths and 16.2% pregnancy terminations).
Results: Among the 583 cases with more than 2 CHD, G1 was D1 in 100% of cases, G5 in 88%. G2 and G10 were almost never D1(Table).
Conclusion: This hierarchical approach using ACC-CHD has proved its utility for clinical and epidemiologic studies, and can provide a structure for various databases.
Table:

ACC-CHD Groups	D1(%)	D2(%)	D3(%)	D4(%)
1. Heterotaxy, including isomerism and mirror-imagery	100	0	0	0
2.Anomalies of the venous return	3	50	30	17
3. Anomalies of the atria and interatrial communications	17	78	5	0
4.Anomalies of the atrioventricular junctions and valves	42	37	16	5
5.Complex anomalies of the atrioventricular connections	88	13	0	0
6.Functionally univentricular hearts	44	34	17	5
7.Ventricular septal defects	52	43	3	1
8. Anomalies of the outflow tracts and VA connections	44	35	15	5
9. Anomalies of the extrapericardial trunks	46	33	16	6
10. Congenital anomalies of the coronary arteries	0	70	30	0