Terminology of Congenital Heart Disease

Glossary and Commentary

One of the fascinations of pediatric cardiology is its complexity. However, this complexity and the various terminologies that strive to express it often baffle interested outsiders — adult cardiologists, pediatricians, internists, radiologists, pathologists, geneticists, embryologists, and referring family physicians — all of whom need and want to understand.

In this issue of Circulation (page 159), Stanger, Rudolph and Edwards¹ present an extensive study of the cardiac malpositions, and they also consider the problem of terminology. Using the segmental approach and segmental terminology, they introduce a notation that uses minimal abbreviation and which therefore should be readily understandable even by those who know relatively little about congenital heart disease. They also indicate that full abbreviation² of the cardiac segments can be helpful to those who are working in this field.

The following brief glossary and commentary are intended to supplement the report of Stanger, Rudolph and Edwards and to try to answer some of the questions of those who are not familiar with the taxonomy of congenital heart disease.

**Cardiac Position:** levocardia (left-sided), dextrocardia (right-sided), mesocardia (centrally located) within the thorax, and ectopia cordis (partially or completely outside the thorax).

**Segmental Approach:**² analysis of the heart in terms of its major developmental units or segments in a step-by-step fashion. The segments are the visceroatrial situs, the ventricular loop, and the conotruncus — or more briefly, the atria, ventricles, and great arteries. Figure 1 is included to assist the reader to visualize what is summarized. Except where otherwise noted, all types of human hearts discussed below are shown in figure 1.

**Chamber Identification:** morphologically or anatomically³ (see Stanger et al.).¹ The morphologically right atrium = RA; the morphologically left atrium = LA; the morphologically right ventricle = RV (Stanger’s fig. 2); and the morphologically left ventricle = LV (Stanger’s fig. 2).

**Situs Solitus:** normal arrangement of viscera and atria, with RA right-sided and LA left-sided. Solitus = usual, ordinary, customary — hence normal.

**Situs Inversus:** inverted arrangement of viscera and atria, with RA left-sided and LA right-sided, as in a mirror-image of situs solitus.

**Situs Ambiguous:**² the ambiguous situs often associated with congenital asplenia, less often with polysplenia, and rarely with a normally formed spleen (not shown in fig. 1). In situs ambiguous, the basic type of visceroatrial situs is anatomically uncertain or indeterminate, because the anatomic findings are ambiguous.

Stanger et al.¹ prefer to break situs ambiguous down into two types of situs:

- **Asplenia:** bilateral right-sidedness, and **Polysplenia:** bilateral left-sidedness.

We regard bilateral right-sidedness and bilateral left-sidedness as helpful mnemonics for teaching, but not as basic biology. For example, the RA is not really bilateral in asplenia, nor is the LA really bilateral in polysplenia.², ⁴, ¹⁰, ¹¹ “Suggestive of” bilateral RA or LA is more accurate, we think. Nonetheless, the distinction between the asplenia and polysplenia syndromes is helpful. Stanger et al. have emphasized the differences between these two syndromes. To balance the picture, it is important to mention the similarities. Hence, we prefer: **Situs ambiguus with asplenia,** and **Situs ambiguous with polysplenia.**

**Heterotaxy Syndrome:** situs ambiguus.

**D-loop:**⁴ RV right-sided relative to LV.

Note that the RV is the right ventricular inflow tract (sinus, or body). The outflow tract is the infundibulum or conus (very different from the true RV anatomically, embryologically and positionally).

Synonyms for D-loop: normally located ventricles, solitus ventricles, noninverted ventricles in situs solitus.

D-loop ventricles in situs inversus are regarded by some as inverted ventricles for situs inversus; others regard them as noninverted ventricles in situs inversus.

In situs ambiguous, D-loop ventricles cannot be described as noninverted or inverted, because the frame of reference, the type of visceroatrial situs, itself is unknown.

D-loop simply says where the RV inflow tract is: it is right-sided (dextro or D) relative to the LV.

**L-loop:**⁴ RV left-sided (levo or L) relative to LV. In situs solitus, L-loop ventricles are inverted. In situs inversus, L-loop ventricles have been regarded as noninverted for situs.

---

From the Children's Hospital Medical Center, Boston, Massachusetts.
Address for reprints: Richard Van Praagh, M.D., The Children's Hospital Medical Center, 300 Longwood Avenue, Boston, Massachusetts 02115.
Received October 11, 1976; revision accepted March 11, 1977.
Figure 1. Diagram of various types of heart, viewed from below. (Reproduced with permission from Van Praagh.*)
inversus, and as inverted — part of situs inversus totalis. In situs ambiguous, L-loop ventricles cannot be described as inverted or noninverted (because the type of visceral atrial situs is unknown).

L-loop simply says where the RV inflow tract is: it is left-sided (levo or L) relative to the LV.

X-loop: type of bulboventricular loop is anatomically uncertain or unknown (X = unknown) (not shown in fig. 1).

Concordant Loop, the ventricular loop is concordant or appropriate relative to the visceral atrial situs: in situs solitus, a D-loop; in situs inversus, an L-loop; and in situs ambiguous, the concordance/discordance concept is not applicable.

Discordant Loop, ventricular loop is discordant or inappropriate relative to the visceral atrial situs: in situs solitus, an L-loop; in situs inversus, a D-loop; and in situs ambiguous, the concept is inapplicable.

Mixed discordant; e.g., mixed levocardia with atrial noninversion means left-sided heart with situs solitus of viscera and atria and discordant or inappropriate ventricles. Since the atria are in situs solitus, the ventricles are inverted (L-loop).

Normally Related Great Arteries: solitus normal and inverted normal — the usual and mirror-image normals, respectively.

Stanger et al. introduced the new terms D-normal (= solitus normal) and L-normal (= inverted normal).

In cardiac malpositions, solitus normally related great arteries do not always have an aortic valve that is to the right of the pulmonary valve; the aortic valve can be relatively left-sided. Hence, D-normal is not always D. But it always is the solitus or usual normal relationship between the great arteries themselves and between the great arteries and the ventricles. For example, the pulmonary artery (PA) always passes to the left of the ascending aorta (Ao), as the great arteries untwist about each other.

Although we prefer the older terms (there is nothing wrong with them), these new ones are generally acceptable synonyms (except in the rare situation mentioned above).

Ventriculo-Arterial Concordance, normally related great arteries. This synonym for normal is accurate, except for anatomically corrected malposition (see below) that also has ventriculo-arterial concordance, but in which the great arteries are not normally interrelated.

Transposition of the Great Arteries: Aorta arising above RV and PA arising above LV. Stanger et al. note that in TGA, the aorta usually is anterior to the PA, but that it can be beside or behind the PA.

D-TGA, aortic valve (AoV) to right (dextro or D) relative to pulmonary valve (PV).

L-TGA, AoV to left (levo or L) relative to PV.

A-TGA, AoV directly anterior (antero or A) relative to PV (not shown in fig. 1).

Complete TGA: physiologically uncorrected TGA; e.g., in situs solitus, usually D-TGA, and in situs inversus, usually L-TGA.

Corrected TGA: physiologically corrected TGA; e.g., in situs solitus, usually L-TGA, and in situs inversus, usually D-TGA.

Noninverted TGA: complete, physiologically uncorrected, and noninverted TGA are all essentially synonymous. In situs solitus, the AoV is normally right-sided.

In D-TGA, the AoV is right-sided. Therefore, D-TGA is a noninverted TGA — there being no right-left reversal of Ao.

In situs inversus, L-TGA is the usual complete, physiologically uncorrected TGA. It may also be regarded as noninverted TGA relative to situs inversus because in situs inversus the Ao is supposed to be left-sided and in L-TGA, it is; i.e., there has been no right-left switching of the Ao relative to what is normal for the situs.

Inverted TGA: physiologically corrected TGA usually is inverted TGA. In situs solitus, L-TGA is inverted TGA. The Ao is to the left; normally in situs solitus, the Ao is to the right. Similarly, in situs inversus, D-TGA may be regarded as inverted or right-left switched. The Ao is to the right; but in situs inversus, the Ao should be to the left.

TGA in Situs Ambiguus: cannot be described accurately as complete (noninverted) or as corrected (inverted) because the visceral atrial situs — the frame of reference — is anatomically indeterminate. Situs ambiguous (not shown in fig. 1) is not a negligible problem. In the series of Stanger et al. it constituted 62% of the cases (40 of 65).

Ventriculo-Arterial Discordance: TGA.

Double Outlet Right Ventricle: Origin of Ao and PA entirely or predominantly above RV.

I agree with Stanger et al. that tetralogy of Fallot should be diagnosed as such and not included in the category of DORV, and that the diagnosis of the Taussig-Bing malformation should be applied only to DORV with subpulmonary ventricular septal defect (VSD), as in the original case.

Double Outlet Left Ventricle (DOLV): origin of Ao and PA entirely or predominantly above LV.

Malposition of the Great Arteries: the great arteries are malposed, but they are not transposed, accurately speaking. Trans = across and ponere = to place. In TGA, both the Ao and the PA are placed across the ventricular septum and so arise above the anatomically inappropriate ventricles.

In malposition of the great arteries (MGA), this is not the case. MGA occurs, for example, with DORV and DOLV.

D-MGA, L-MGA and A-MGA: MGA with the AoV to the right (dextro or D), to the left (levo or L), or anterior (antero or A) relative to the PV, respectively.

Single (Common) Ventricle: When there is no identifiable ventricular septum — single RV (type B), absent or rudimentary ventricular septum (type C) and morphologically unidentified ventricular myocardium (type D) — MGA applies accurately, but TGA does not.

However, in the most frequent form of single ventricle, single LV* with infundibular outlet chamber (type A) (double inlet LV), TGA does apply accurately because a malformed ventricular septum is present which is well developed on the LV side but virtually absent on the RV side (because the RV is absent).

Anatomically Corrected Malposition (ACM): malposed Ao nonetheless above LV and malposed PA nonetheless above RV. In this sense, the malposition of the great arteries is anatomically corrected.

Notation

The segmental terms may be written out in full, or minimally abbreviated, or fully abbreviated. In the following examples, please see figure 1.
Normal Heart

No abbreviation: situs solitus of viscera and atria, D-loop, solitus normally related great arteries.
Minimal abbreviation: situs solitus/L-loop/D-trans.
Full abbreviation: {S,D,S}.
S = situs solitus; D = D-loop; S = situs solitus normal great arteries. Braces \{ \} indicate a set or subset. The cardiac segments are members of the set or subset and are separated by commas. Braces and commas are standard set notation. (Braces are readily available for typewriters and hence do not need to be written in by hand.)

Inverted Normal Heart

No abbreviation: situs inversus of viscera and atria, L-loop, inverted normally related great arteries.
Minimal abbreviation: situs inversus/L-loop/L-trans.
Full abbreviation: {I,L,L}.

Complete Transposition in Situs Solitus

No abbreviation: transposition of the great arteries with situs solitus of viscera and atria, D-loop, and D-transposition.
Full abbreviation: TGA {S,D,D}.

Corrected Transposition in Situs Inversus

No abbreviation: transposition of the great arteries with situs inversus of viscera and atria, D-loop, and D-transposition.
Full abbreviation: TGA {I,D,D}.

Taussig-Bing Malformation in Situs Solitus

No abbreviation: double outlet right ventricle with situs solitus of viscera and atria, D-loop, and D-malposition of the great arteries, and subpulmonary ventricular septal defect.
Minimal abbreviation: situs solitus/D-loop/D-DORV with subpulmonary VSD.
Full abbreviation: DORV {S,D,D} with subpulmonary VSD.

DORV {S,D,D} is shown in figure 1, but without associated malformations (such as VSD). In the next two examples, associated malformations (not shown in fig. 1) will be included to indicate how they are incorporated in order to make a complete anatomic diagnosis. (Physiologic diagnoses are also crucially important, of course, but are beyond the scope of this editorial.)

Double Outlet Right Ventricle with Aorta to the Left, etc. (not shown in fig. 1)

No abbreviation: double outlet right ventricle with situs solitus of viscera and atria, D-loop, L-malposition of the great arteries, subaortic ventricular septal defect and pulmonary outflow tract stenosis.
Minimal abbreviation: situs solitus/D-loop/L-DORV with subaortic VSD and PS.
Full abbreviation: DORV {S,D,L} with subaortic VSD and PS.

Dextrocardia with Corrected Transposition, etc.

No abbreviation: dextrocardia with transposition of the great arteries, situs solitus of the viscera and atria, L-loop, L-transposition, subpulmonary ventricular septal defect, bilateral conus, pulmonary outflow tract stenosis and regurgitation of the left-sided tricuspid valve.
Minimal abbreviation: situs solitus/L-loop (D)/L-trans with subpulmonary VSD, bilateral conus, PS and TR (left-sided).
Full abbreviation: TGA {S,L,L} with dextrocardia, subpulmonary VSD, bilateral conus, PS and TR (L).

Conclusion

Diagnoses in congenital heart disease should be anatomic, specific, and not overlapping. A step-by-step or segment-by-segment approach to diagnosis is very helpful in complex congenital heart disease,17 as the extensive yet precise report of Stanger, Rudolph and Edwards1 illustrates.

It is hoped that this brief editorial, the references, and the paper of Stanger et al.1 will help to make complex congenital heart disease easier to understand and to describe.

Richard Van Praagh, M.D.
References


Insurability and Employability

Congenital Heart Disease and Innocent Murmurs

"CAN I GET A JOB when they see this scar on my chest?"
"I feel OK now. How's it going to help me to have this septal defect closed?" "Doctor, if you say my child has an innocent murmur, will that be bad for his record later on when he applies for a job or wants to get insurance?"

Every day some physician hears a question such as this. Each year about 20,000 Americans born with a cardiac defect enter the adult world. Over half of the rest of the population becoming adults have had an innocent or functional murmur, since such a murmur is so prevalent in childhood. While these people have been growing up, so have the fields of pediatric cardiology and cardiovascular surgery. Medical students have been trained to listen carefully as physicians and to record heart sounds and murmurs. At the same time that the natural history of the common kinds of congenital heart diseases is being documented,1,2 it is being rewritten by the continuing advances in diagnostic capabilities as well as in medical and surgical treatment. Analyses of long-term results of that treatment are appearing each month in some journal devoted to cardiology or to cardiovascular surgery, and modifications of previous recommendations are being advised. Meanwhile, amid these changing circumstances, insurance companies find themselves with little actuarial data on congenital heart disease, unlike their experience with hypertension, coronary artery disease and rheumatic heart disease where such data are fairly abundant.3

How does the physician interested in his patient's long-term well-being and productivity as an adult guide the child with a congenital cardiac defect to be competitive in the job pool and to assume the other responsibilities of the adult in today's society, including insurance for himself and his family? An even more common question, because of the larger numbers involved, is what will be the effect on later insurability and employability of faithfully recording on the normal child's report that there is a midprecordial, mid-systolic, vibratory murmur, grade II out of VI in intensity, with physiologic splitting of the second heart sound?

Insurability is an overt expression of acceptance by society of the prospect of a relatively normal life pattern.4,5 As such, it is important to many normal individuals, and it has particular meaning to those with a cardiac defect that may or may not have required surgery in the past, and that may or may not need cardiac surgery in the future.

In order to gain an understanding of the problems of clinical cardiology and of insurance medicine in evaluating patients with congenital cardiac defects or with innocent murmurs and to modify opinions as experience and additional information become available,4 the American Heart Association together with the Association of Life Insurance Medical Directors of America arranged a series of conferences. The first was in May of 1963,4 the second in May of 1965,4 and the third in June of 1972. The deliberations of that last meeting, which included representatives of the Industrial Medicine Association as well, have since been reviewed, updated and approved by the physicians representing the Life Insurance Medical Directors and Industrial Medicine and by the Council on Cardiovascular Disease in the Young of the American Heart Association. The names of the participants, who concurred in the final version of the report which appears in the News from the American Heart Association section in this issue of Circulation are as follows:

R Van Praagh

Circulation. 1977;56:139-143
doi: 10.1161/01.CIR.56.2.139
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1977 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/56/2/139.citation

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/