MEDICAL SCIENCE CARDIOLOGY

# Standardizing Congenital Heart Disease Nomenclature: A Comprehensive Analysis of Van Praagh and Anderson Approaches

Published: September 2, 2026 | Index ID: #340

The diagnosis and surgical management of congenital heart disease (CHD) represent some of the most complex challenges in modern medicine. At the heart of these challenges lies a fundamental requirement: a precise, unambiguous language to describe cardiac malformations. The **Atlas of Congenital Heart Disease Nomenclature**, specifically the work curated by William B. Kyle and influenced by pioneers like Richard Van Praagh and Robert H. Anderson, serves as the definitive textual bridge between disparate morphological philosophies. For clinicians, surgeons, and pathologists, understanding the nuances of these nomenclature systems is not merely an academic exercise but a critical component of perioperative safety and diagnostic accuracy.

## The Critical Need for Standardized Nomenclature in Pediatric Cardiology

Before the mid-20th century, the description of congenital cardiac defects was often idiosyncratic, relying on eponyms or vague descriptive terms that varied by institution. As cardiac surgery advanced, particularly with the advent of cardiopulmonary bypass, the need for a rigorous, reproducible method of describing the heart became paramount. Miscommunication regarding the position of a ventricle or the connection of a great artery could lead to catastrophic surgical errors.

Today, the medical community primarily utilizes two major frameworks for cardiac description: the **Segmental Approach** (championed by Van Praagh) and **Sequential Segmental Analysis**(championed by Anderson). While they share a common goal of logical anatomical description, their methodologies, notations, and philosophical underpinnings differ significantly. This article provides a deep dive into these systems, exploring how they categorize the infinitely variable landscape of the malformed heart.

## Theoretical Framework: The Building Blocks of Cardiac Morphology

To understand the nomenclature systems, one must first master the concept of the **morphological method**. This principle dictates that a cardiac chamber is defined by its intrinsic anatomical features rather than its position in the chest or its connections to other chambers. In CHD, the heart is viewed as a series of three primary segments:

- The Atria: Defined by the morphology of the auricles (appendages).
- The Ventricles: Defined by the pattern of apical trabeculations and the presence or absence of a tricuspid or mitral valve.
- The Great Arteries: Defined by their branching patterns and ultimate destination (lungs vs. systemic circulation).

### Morphological Indicators for Chamber Identification

In the **Van Praagh and Anderson** approaches, identification begins with specific markers:

- Morphological Right Atrium (RA): Characterized by a broad-based, blunt-shaped appendage and the presence of the crista terminalis and pectinate muscles that extend around the entire vestibule of the atrioventricular valve.

- Morphological Left Atrium (LA): Characterized by a narrow, finger-like, crenulated appendage and the absence of a crista terminalis.
- Morphological Right Ventricle (RV): Distinguished by coarse apical trabeculations, the presence of a moderator band, and a tricuspid valve that is more apically displaced than the mitral valve.
- Morphological Left Ventricle (LV): Distinguished by fine, "suit-and-tie" or "criss-cross" trabeculations and a mitral valve with two primary papillary muscles.

## The Van Praagh Approach: The {S, D, S} Notation System

Developed at Boston Children's Hospital, the Van Praagh system is renowned for its concise, symbolic notation. It views the heart through the lens of embryological development, specifically focusing on the concept of "situs" and "looping."

### The Three-Segment Notation

The hallmark of the Van Praagh method is the use of a triplet in curly brackets, such as **{S, D, S}**. Each position in the triplet represents a specific segment:

1. First Letter (Atrial Situs): Represents the arrangement of the atria and viscera. S (Situs Solitus - normal), I (Situs Inversus - mirror image), or A (Situs Ambiguus/Heterotaxy).
2. Second Letter (Ventricular Loop): Describes the direction of the bulboventricular loop during development. D (Dextro-loop, resulting in a normal RV to the right of the LV) or L (Levo-loop, resulting in an inverted arrangement).
3. Third Letter (Great Artery Relationship): Describes the position of the aortic valve relative to the pulmonary valve. S (Solitus/Normal), I (Inverted), D (D-transposition), or L (L-transposition).

### Mathematical Logic of the Van Praagh System

The power of the Van Praagh system lies in its predictive capability. For instance, in a **concordant** heart, atrial situs (S) and ventricular loop (D) usually align. When they do not—such as in an {S, L, L} heart—the clinician immediately recognizes a **Congenitally Corrected Transposition of the Great Arteries (ccTGA)**, where the blood flows in the correct physiological sequence but through the "wrong" anatomical chambers.

## The Anderson Approach: Sequential Segmental Analysis

In contrast to the symbolic shorthand of Van Praagh, the **Anderson approach** (widely adopted in the UK and Europe) emphasizes a descriptive, step-by-step prose analysis. This method focuses heavily on the **connections** between segments rather than just their spatial relationships.

### The Four-Step Diagnostic Process

Anderson's methodology requires the clinician to define the heart by answering four sequential questions:

1. What is the atrial arrangement? (Solitus, Inversus, or right/left isomerism).
2. How are the atria connected to the ventricles? (Atrioventricular connection: Concordant, Discordant, Ambiguous, Univentricular).
3. What is the ventricular topology? (Right-hand vs. Left-hand pattern).
4. How are the ventricles connected to the great arteries? (Ventriculo-arterial connection: Concordant, Discordant, Double Outlet, Single Outlet).

### Terminology Differences: Connections vs. Relations

One of the primary distinctions in Anderson's work is the insistence on separating "connections" from "relations." A connection describes which chamber flows into which, whereas a relation describes where they sit in space. This

distinction is vital in complex pathologies like **Double Outlet Right Ventricle (DORV)**, where the connection is the defining feature, regardless of the spatial rotation of the great vessels.

## Comparative Analysis: Van Praagh vs. Anderson

While both systems are technically accurate, they use different lenses to view the same pathology. The following table highlights the core differences in their descriptive philosophies.

| Feature | Van Praagh Approach | Anderson Approach |
|---|---|---|
| Primary Focus | Embryological development and spatial shorthand. | Step-by-step anatomical connections. |
| Notation Style | Symbolic triplets (e.g., {S, D, S}). | Descriptive prose/Sequential Analysis. |
| Atrial Identification | Focuses on Visceral Situs. | Focuses on Isomerism (Appendage morphology). |
| Ventricular Description | Uses "Looping" (D-loop vs. L-loop). | Uses "Topology" (Right-hand vs. Left-hand). |
| Great Vessel Terms | D-Transposition / L-Transposition. | Discordant Ventriculo-arterial Connection. |
| Complexity Handling | High; requires mastery of the symbolic code. | High; requires detailed descriptive accuracy. |

## Technical Analysis of Ventricular Topology

A central concept in both systems, though named differently, is the spatial orientation of the ventricles. In the Van Praagh system, this is the **Ventricular Loop**. In the Anderson system, this is **Topology**.

### The Right-Hand Rule of Ventricular Topology

Anderson introduced a practical heuristic known as the "Hand Rule" to determine ventricular topology, which is essential for understanding the internal architecture of the heart:

- Right-Hand Topology (D-Loop): If you place your right hand on the septal surface of the morphological right ventricle such that your thumb is in the tricuspid valve (inlet) and your fingers point toward the pulmonary valve (outlet), and the palm rests on the septum, it is a right-hand pattern.
- Left-Hand Topology (L-Loop): If the left hand fits this configuration better, it is a left-hand pattern (typically found in corrected transposition).

This technical distinction is crucial for surgeons planning a **Septal Defect** repair, as the location of the **Conduction System**(the Bundle of His) varies predictably based on the topology. In right-hand topology, the bundle typically runs along the postero-inferior rim of a VSD; in left-hand topology, it is often antero-superior.

## Clinical Case Study: Describing Transposition of the Great Arteries (TGA)

To illustrate the practical application of these nomenclature systems, let us examine a standard case of **D-TGA** (Complete Transposition).

### The Van Praagh Description: {S, D, D}

In this notation, the surgeon immediately knows:

- S: Atrial situs is normal (Situs Solitus).
- D: The ventricles looped to the right (Normal ventricular positions).
- D: The aorta is to the right (Dextro) and anterior of the pulmonary artery, and it arises from the RV (Transposed).

### The Anderson Description

The report would state: "Atrial situs solitus, concordant atrioventricular connections, discordant ventriculo-arterial connections, with the aorta arising from the morphological right ventricle and the pulmonary artery from the morphological left ventricle."

### Analysis of Utility

The Van Praagh system provides a rapid-fire summary for experienced clinicians, while the Anderson system ensures that no detail of the connection is lost to a reader who may not be familiar with the shorthand. The **Atlas of Congenital Heart Disease Nomenclature** advocates for a synthesis of these views to ensure maximum clarity.

## Operational Challenges and Error Mitigation in the Field

Despite these sophisticated systems, errors in CHD nomenclature persist, often due to "hybridization"—the mixing of terms from different systems without proper definition. Common failure modes include:

1. Eponymous Ambiguity: Using terms like "Taussig-Bing Heart" instead of "DORV with subpulmonary VSD." Eponyms do not describe the anatomy and can be misinterpreted.
2. Misidentifying Situs: In cases of Heterotaxy, assuming that the liver position dictates the atrial position. Both Van Praagh and Anderson emphasize looking at the atrial appendages themselves.
3. Confusing Relations with Connections: Describing a vessel as "transposed" just because it is anterior, even if it is connected to the correct ventricle (a condition known as Malposed Great Arteries).

### The Solution: The International Pediatric and Congenital Cardiac Code (IPCCC)

To resolve these conflicts, the **International Pediatric and Congenital Cardiac Code (IPCCC)** was established. It acts as a standardized dictionary that maps Van Praagh's symbols to Anderson's descriptions, providing a unified language for the **ICD-11**and global surgical databases. The William B. Kyle atlas is a vital tool in teaching clinicians how to map their local findings to this global standard.

## Advanced Procedural Execution: A Step-by-Step Guide to Cardiac Categorization

For a technologist or fellow performing an echocardiogram or reviewing a cardiac MRI, the following procedural workflow is recommended to ensure accurate nomenclature application:

### Step 1: Determine Visceral and Atrial Situs

Locate the liver, stomach, and spleen. Use high-resolution imaging to visualize the **morphology of the atrial appendages**. Do not rely on the venous connections (e.g., IVC position) alone, as these can be anomalous in heterotaxy.

### Step 2: Identify Ventricular Morphology and Topology

Look for the **moderator band**and the apical insertion of the tricuspid valve to identify the RV. Use the "Hand Rule" to determine if the topology is D-loop or L-loop. Note the presence of any ventricular septal defects (VSDs) and their relationship to the outflow tracts.

### Step 3: Evaluate Atrioventricular (AV) Connections

Determine if each atrium is connected to a ventricle. Identify if there are two valves (mitral and tricuspid), a common AV valve, or if one valve is **atretic**(absent). This step defines whether the heart is biventricular or univentricular in its connection pattern.

### Step 4: Evaluate Ventriculo-arterial (VA) Connections

Track the origin of the Aorta and the Pulmonary Artery. Are they coming from the correct ventricles (concordant), the wrong ones (discordant), or both from one (Double Outlet)?

### Step 5: Document Spatial Relationships

Finally, describe the spatial orientation of the great arteries (e.g., D-malposition, L-malposition, side-by-side) to assist the surgeon in planning the incision and vessel mobilization.

## Implications for 3D Modeling and Future Diagnostics

As we move into an era of 3D printing and virtual reality (VR) surgical planning, the importance of nomenclature is shifting from 2D descriptions to 3D spatial data. However, the **Segmental Approach**remains the underlying logic for 3D segmentation. A computer algorithm must be told which voxels represent the "morphological RV" based on the same trabecular patterns defined by Van Praagh and Anderson decades ago.

Furthermore, the integration of artificial intelligence (AI) in echocardiography relies on large, correctly labeled datasets. If the input data is labeled inconsistently (using a mix of poorly defined Anderson and Van Praagh terms), the AI's predictive power for surgical outcomes is diminished. Thus, the rigorous study of nomenclature as presented in the **Atlas of Congenital Heart Disease Nomenclature**is more relevant today than it was at its inception.

The study of cardiac nomenclature is the study of the heart's very identity. Whether one prefers the algebraic elegance of Van Praagh’s triplets or the meticulous descriptive sequence of Anderson’s analysis, the objective remains the same: to provide the most accurate possible map of a child’s heart. By bridging these two systems, the work of William B. Kyle and his colleagues ensures that no matter where a surgeon is trained, the language of the operating room remains a universal constant, dedicated to the precision and safety of pediatric cardiac care.

**Tags: #Congenital Heart Disease #Cardiac Nomenclature #Van Praagh Approach #Anderson Sequential Analysis #Pediatric Cardiology**