A Question of Definition

What is a Ventricle?
The Single-Ventricle Trap
Richard Van Praagh, Ivan David, Stella Van Praagh
Children’s Hospital Medical Center, Boston, Massachusetts

SUMMARY. Whether a chamber is or is not a ventricle is determined by its myocardial morphological characteristics, not by the entering atrioventricular (AV) valves. The anatomic characteristics of the morphologically right ventricle and the morphologically left ventricle have been well described, are widely understood, and do not need to be changed. What a ventricle is should not be redefined in terms of the AV connections because such redefinition is based on the classic definition of single ventricle, which itself is not satisfactory. This old definition is wrong in principle, violates the morphological method of diagnosis and designation of the cardiac chambers, and has two kinds of exceptions. Consequently the classic definition of single ventricle must not be used as a paradigm (model) for the redefinition of what a ventricle is or is not. The approach and terminology based on the use of the unsatisfactory single-ventricle paradigm should be discontinued, eg, “primitive ventricle,” “main chamber,” “accessory chamber,” “trabecular pouch.” Being unnecessary, these nonmorphological components of the terminology of congenital heart disease may be omitted. Morphological anatomy, unadorned, is the key to clarity, simplicity, and accuracy.

KEY WORDS: Definition of the ventricles — Morphological method of diagnosis and naming of the ventricles — Single ventricle — Tricuspid atresia — Mitral atresia — Straddling atrioventricular valves

Oddly enough the question “What is a ventricle?” recently has become controversial. How did this happen, and why?

In telling this tale we are deliberately going to avoid pointing fingers and naming names, not only because all of the actors in this drama are personal friends and respected colleagues, but also because the real villain of the piece turns out not to be any of our colleagues, but rather is the classic definition of single ventricle: Single ventricle is present when both atrioventricular (AV) valves or a common AV valve open(s) into one ventricular chamber [7, 8].

Although we accepted this old definition as the starting point of our initial study of single ventricle [7], application of the morphological method of diagnosis and naming of the cardiac chambers, introduced by Lev [4] in 1954, revealed that the classic definition of single ventricle is far from satisfactory. The morphological method may be stated as follows [4, 7, 8]:

1. Cardiac chambers are diagnosed and named in terms of their gross myocardial morphological characteristics.

2. Cardiac chambers are not diagnosed and named in terms of the vessel or valve of entry or exit, nor in terms of relative position (such as right sided or left sided), nor in terms of the type of blood conveyed by the chamber (arterial or venous), because these are variables in congenital heart disease.

The classic definition of single ventricle is an unwitting violation of the morphological method...
because this old definition attempts to state whether a single ventricle is present by means of the AV valves, which is specifically prohibited by the morphological method.

Since the classic definition of single ventricle is not consistent with the morphological method, it is not surprising that morphological anatomic analysis—based on an understanding of the gross characteristics of the morphologically right ventricle (RV) and of the morphologically left ventricle (LV)—reveals that there are two types of exception to this old definition. These exceptions are (1) the "single" RV problem and (2) the single LV problem.

The 'Single' RV Problem

When you find a case in which both AV valves or a common AV valve open(s) entirely or predominantly into a large RV, and there is also a small LV that receives less than half of an AV valve, you have essentially two choices:

1. You may say, "This case satisfies the general definition of single ventricle. However, in terms of morphological anatomy, a small LV is present. Therefore, in the interests of morphological anatomic accuracy, such cases should be excluded from the diagnostic category of single ventricle."

   We think this is the correct choice. Whenever there is a conflict between the results of morphological anatomic analysis and findings based on a classic definition, the results of morphological anatomic analysis should be preferred because old definitions may not be infallible. They may prove to have important exceptions, and they may even be inconsistent with modern methods of analysis (such as the morphological method), as we have seen.

   Morphological anatomic analysis shows that double-inlet RV and common-inlet RV may or may not have single RV [8]. Usually a small LV is present. Accurately speaking, such cases have biventricular hearts with a large RV and a small LV, not univentricular hearts with a large RV and no LV. Rarely, however, cases with double-inlet or common-inlet RV may have single RV, when no LV is demonstrable [7, 8].

   Since the classic definition of single ventricle is nonmorphological, it fails to exclude cases with a small or rudimentary LV. This is why the old definition of single ventricle needs to be supplemented by morphological anatomic analysis—to ensure anatomic accuracy.

2. The other choice is to say, "This is a single ventricle by definition. The chamber that looks like a small LV therefore cannot be regarded as a small LV. We must call it something else. Otherwise we will not be able to regard this as a single RV, ie, as a univentricular heart of RV type. If we say that a small LV is present, then logically we must consider this to be a biventricular heart. But this is a univentricular heart by definition. The definition of single ventricle (univentricular heart) therefore requires us to call this small chamber of LV morphology something other than 'a small LV.' Let us call it an 'accessory chamber'—an 'outlet chamber' if there is an arterial outlet, or a 'trabecular pouch' if there is no outlet. The definition of single ventricle (univentricular heart) also forces us to redefine what a 'ventricle' is. To be consistent with the definition of single ventricle, we must say that to be called a 'ventricle,' a chamber must receive 50% or more of an AV valve."

   The definition of single ventricle is a trap. To get caught, the only mistake you need to make is to assume that this definition is satisfactory and therefore should be applied in all relevant situations. If you make this one assumption—which we know to be erroneous based on morphological anatomic analysis—logic then "forces" you to make other derivative errors.

   Morphological anatomic analysis and logic both indicate that:

   large RV + small LV ≠ single RV

   The real reason for the redefinition of ventricle is failure to understand the limitations of the classic definition of single ventricle. This redefinition is often presented as having either an anatomic or an embryologic basis. It is true that a ventricle normally has an inlet zone, including the AV connection, a trabecular zone, and an outlet zone. It does not follow that if a chamber at the ventricular level receives only 49% of an AV valve (instead of 51% or more), that such a chamber is not a ventricle. It does follow that such a chamber is not a normal ventricle. But there is no anatomic or embryologic reason to assert that such a chamber is not a ventricle at all. The real reason underlying this otherwise incomprehensible assertion is the unsatisfactory old definition of single ventricle.

   The names of the ventricles should not change, depending on the degree of straddling of an AV valve. This is not a practical approach. Moreover, it violates a principle of logic:

   One variable (such as the ventricular anatomy) should not be defined primarily in terms of another variable (such as the anatomy of the AV valves). Each variable (such as the anatomy of the ventricles) should be defined primarily in terms of itself, ie, primarily in terms of its own myocardial morphological characteristics. This is what the morphological method does [4, 7, 8].