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Original Article

The univentricular heart: Revisited

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ABSTRACT

Introduction: A univentricular (UV) heart is found when both atria drain mainly into one ventricle; while if either ventricle cannot sustain the systemic or the pulmonary circulation, a *functionally* UV heart emerges. UV heart is the source of hot debates and discussions for decades that stem from controversies in terminologies, analysis, and management.
Objective: To study the morphological pattern of univentricular hearts.
Design: Cohort, descriptive study.
Setting: Review 13-year data from the file system of a high-volume, tertiary, teaching cardiac centre and study the patients having pathologies meeting the definition of UV heart.
Results: 461 patients have UV hearts; constituting 1.4% of all patients with a congenital heart disease (CHD) visiting the centre of the study. *Functionally* UV heart in the form of biventricular atrioventricular (AV) connection plus one ventriculoarterial (VA) connection was seen in 179 cases (38%) with aortic atresia being the most common (*N* = 98; 55%). On the other hand, UV AV connection was seen in 282 cases (62%). Under this category, 203 cases (72%) show an absent AV connection and 79 hearts (28%) with double-inlet anatomy.
Conclusion: UV heart is a rare complex CHD with lots of scientific debates. It is hoped that this long-term study will be the basis and a reference to paediatric cardiologists, echocardiographers, and other interested professionals.

El corazón univentricular: nueva revisión

Palabras clave:
Corazón univentricular
Ventrículo único
Ventrículo común
Reparación de 2 ventrículos
Reparación de un ventrículo

RESUMEN

Introducción: Un corazón univentricular (UV) se encuentra cuando ambas aurículas drenan principalmente en un ventrículo; mientras que, si el ventrículo no puede sostener la circulación sistémica o pulmonar, emerge un corazón funcionalmente UV. El corazón UV es la fuente de acalorados debates y discusiones durante décadas que derivan de controversias en terminología, análisis y gestión.
Objetivo: Estudiar el patrón morfológico de los corazones UV.
Diseño: Estudio descriptivo de cohorte.
Escenario: Revisar los datos de 13 años del sistema de archivos de un centro cardíaco docente de alto volumen y terciario, y estudiar a los pacientes con enfermedades que cumplen con la definición de corazón UV.
Resultados: Cuatrocientos sesenta y un pacientes tienen corazones UV; constituye el 1,4% de todos los pacientes con cardiopatía congénita (congenital heart disease [CHD]) que visitan el centro del estudio. El corazón funcionalmente UV en forma de conexión biventricular auriculoventricular (AV) más una conexión ventriculoarterial (VA) se observó en 179 casos (38%) y la atresia aórtica fue la más común (N = 98; 55%). Por otro lado, la conexión UV/AV se observó en 282 casos (62%). En esta categoría, 203 casos (72%) muestran una conexión AV ausente y 79 corazones (28%) con anatomía de doble entrada.
Conclusión: El corazón UV es una rara CHD, además de compleja, con muchos debates científicos. Se espera que este estudio a largo plazo sea la base y una referencia para los cardiólogos, ecocardiografistas y otros profesionales interesados en pediatría.

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Introduction

The segmental approach, applied for the analysis of the cardiac malformation, has improved the diagnosis and categorization of the complex congenital heart disease (CHD).

Van Praagh[1–3] first highlighted the merits to consider the malformed heart in terms of arterial, ventricular, and atrial components. Some may regard the heart as a "three-floor-complex" – with the ventricles as the first floor, aorta and pulmonary artery (PA) as the second one, and the atria being the platform – connected together through valve orifices and divided by septa.[4,5]

The three segments are joined by two connections, called atrioventricular (AV) and ventriculoarterial (VA). AV junction connects the atria with their corresponding ventricles and the VA one connects the ventricles with their great arteries. In a normal heart, each junction has two different valves: tricuspid and mitral at the AV and aortic and pulmonary in the VA junction.[4]

The AV connection is said to be *biventricular* if each atrium joins on ventricle. When the right atrium (RA) joins the right ventricle (RV), and the left atrium (LA) with the left ventricle (LV), the connection is termed *concordant*. Further, a *discordant* connection is found when the RA joins the LV, and the LA is connected to the RV.[4]

However, sometimes the AV connection cannot be determined as discordant or concordant in the presence of left or right atrial isomerism. In this setting, the topological configuration of the ventricles (i.e. L- or D-loop) must be added.[6]

A *univentricular* heart is found when both atria are connected mainly with one ventricle due to one of two possibilities: 1. *Double-inlet connection*: the presence of single common AV valve or two separate valves that drain predominantly into one ventricle. 2. *Absent connection*: one of the AV valves is atretic or absent. This leads to the formation of two different ventricles, one is small and hypoplastic and the other is of good size. The latter could have left or right morphological pattern or, on rare occasions, *indeterminate*. Determining the type of the ventricular loop is an essential step because an absent AV connection might be seen with a discordant loop.

At the VA connection, the anatomy is *discordant* when the aorta arises from the RV and the PA from the LV, or *concordant* when the great arteries take origin from their corresponding ventricles.[6]

Double-outlet VA connection may be seen with one and a half (or more) of the great vessels originate from one ventricle. The *single-outlet* connection is present when only one patent great vessel arises from the heart, e.g. single pulmonary outlet with atretic aortic valve, a single aortic outlet with atretic pulmonary valve, or single outlet with persistent truncus arteriosus (PTA).

The *univentricular* hearts are, therefore, the result of univentricular AV connections with resulting one large and another hypoplastic ventricle. Nevertheless, there are hearts with normal AV and VA connections in which one ventricle cannot sustain the systemic or the pulmonary circulation. This is the case of aortic stenosis/atresia causing hypoplastic LV or pulmonary stenosis/atresia, or with Ebstein malformation of the tricuspid valve leading to hypoplastic RV.

Many researchers[7–11] unified different anatomical cardiac malformation under the term *functionally univentricular heart* which is characterised by the fact that either ventricle cannot sustain the systemic or the pulmonary circuit because of its diminutive size or deficient function. In this scenario, the heart is not a candidate for biventricular repair; instead, single-ventricle or one-and-half ventricle repair is feasible.

In this study, we reviewed our 13-year data collection with the aim to identify the morphological characteristics of the hearts fulfilling the new concept of *univentricular* heart.

Patients and methods

In this "descriptive cohort" study, we reviewed the file system of a high-volume, busy, tertiary, teaching cardiac centre in Baghdad-Iraq (namely, Ibn Al-Bittar Centre for Cardiac Surgery). We examined all the files made between January 2004 and December 2016 (i.e. 13 years) and identified the patients having cardiac malformations corresponding to the definition of *univentricular* hearts. A univentricular heart is defined by the fact that both atria are connected mainly with one ventricle due to one of two possibilities: double-inlet or absent connection.[6] Cases with a *functionally* univentricular heart were included. The latter is identified when either ventricle cannot sustain the systemic or the pulmonary circuit because of its diminutive size or deficient function.[7–11]

For each patient, we recorded the following echocardiographic findings (when available): cardiac position, atrial situs, the morphology of the ventricles, great arteries anatomy and relation, AV and VA connections, and associated anomalies. Each ventricle is regarded as having left, right or indeterminate morphology and being hypoplastic or dominant depending on its size. D- and L-looping of the ventricles were identified. D-loop topology represents a morphological RV in a right-anterior location and the morphological LV in a left-posterior location. On the other hand, L-loop is found when the morphological RV is in the left-anterior and the LV in a right-posterior position.

The nature of the AV valves was identified; whether there is two patent valves, single (common) valve, atretic valve, overriding or straddling valves. Straddling is present when the tensor apparatus (i.e. chordae tendineae and papillary muscles) is inserted – through a VSD – into the contralateral ventricle. While overriding is considered to be present when there is a commitment of the annulus of the AV valve to two ventricular chambers.[12]

Echocardiography was done using GE Vivid 3 machine (GE Healthcare, USA) in the period between 2004 and 2010; then GE Vivid E9 machine (GE Healthcare, USA) for the remaining period of the study. Two kinds of ultrasonic transducers were used – 3.0 and 5.0 MHz.

We reviewed cardiac catheterization data (when available) for those patients with univentricular hearts. Haemodynamics, O_2 saturation, and angiographic information were recorded. Different operators have performed the echocardiographic and catheterization studies.

No necropsy (pathological) specimens have been included in this study.

Results

During the period of interest (2004–2016), we found 31,205 newly-diagnosed cases with CHD visited the centre of the study. Cases that met the definition of the univentricular heart were seen in 461 patients (1.4%).

We identified two sets of hearts: first, those with biventricular AV connection plus one VA connection. Second, hearts with univentricular AV connection.

Biventricular AV connection plus one VA connection: 179 cases

In this study, 98 patients (55%) were found having atresia of the aortic valve with intact ventricular septum (IVS), and 81 (45%) with atresia of the pulmonary valve with IVS. Patients with VSD and either aortic (143 cases) or pulmonary atresia (232 cases) were excluded because they often show good-sized ventricles.

All cases of aortic or pulmonary atresia have concordant AV connections and situs solitus.