ECHOCARDIOGRAPHIC MORPHOLOGICAL CLASSIFICATION OF UNIVENTRICULAR HEART PATIENTS IN SULAIMANI PEDIATRIC TEACHING HOSPITAL / KURDISTAN / IRAQ

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ABSTRACT

Background

A "single ventricle" or "univentricular heart" was characterized by lacking two well-developed ventricles, which thereby excluded hearts with nonseptable but well-formed ventricles. The proposed definition of univentricular heart encompassed double-inlet atrioventricular connection, absence of atrioventricular connection (mitral or tricuspid atresia), double-outlet right ventricle, common atrioventricular valve with only one well-developed ventricle, and heterotaxy syndrome (single ventricle heterotaxy syndrome).

Objectives

To determine the echocardiographic morphological classification of patients with univentricular heart disease and the subclassification of each type in Sulaimani PediatricTeaching Hospital-Cardiology Unit–Kurdistan-Iraq.

Patients and Methods

A retrospective study conducted in pediatric teaching hospital in Suaimani city from September 2009 to January 2014. All children with the confirmed diagnosis of single ventricle were included in the study, hypoplastic left heart syndrome have been excluded from the study. The source of information was the medical and echocardiography records from cardiology unit of the pediatric teaching hospital in Sulaimani by the same pediatric cardiologist as an operator.

Results

Among 100 cases, 73 cases had left ventricle morphology, 24 cases had right ventricle morphology, and 3 cases of indeterminate type. 50 cases were tricuspid atresia, 22 cases were double inlet left ventricle, 17 cases were double outlet right ventricle, 10 cases were common atrioventricular valve, and one case was double inlet right ventricle. Type (I) tricuspid atersia was the most common subtype of tricuspid atresia (80%). Transposition of great arteries was the most common type of double inlet left ventricle (95%). Tetrallogy of Fallot was the most common subtype of double outlet right ventricle (59%). Ventricular septal defect was the most common associated anomaly, followed by atrial septal defect.

Conclusion

Tricuspid atresia is the most common type of univentricular heart (with exclusion of hypoplastic left heart synsdrome). Tricuspid atersia with normally related great arteries is the most common subtype of tricuspid atersia. Right isomerism is more common in male patients, and right isomerism is more common than left isomerism. Ventricular septal defect & atrial septal defect are the most common associated anomalies. There is a possibility of genetic role in patients with tricuspid atersia that increases its incidence in other siblings.

Keywords: *Tricuspid atresia; Echocardiography; Single ventricle.*

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INTRODUCTION

A “single ventricle” was characterized by lacking 2 well-developed ventricles, which thereby excluded hearts with nonseptable but wellformed ventricles. Hypoplastic left heart syndrome was recognized as a common form of univentricular heart but was classified independently [1].

The proposed definition of univentricular heart encompassed double inlet AV connections [double inlet left ventricle or right ventricle], absence of AV(atrioventricular) connection (mitral or tricuspid atresia), double outlet right ventricle, common AV valve and only 1 well-developed ventricle (unbalanced common AV canal defect), and heterotaxy syndrome (single ventricle heterotaxy syndrome) [2].

Heterotaxy syndromes refer to disorders of lateralization whereby the arrangement of abdominal and thoracic viscera differ from normal and mirror-image of normal. Van Praagh et al. have emphasized that it is inaccurate to label hearts in this category as “single ventricle”, given that there are always two ventricular chambers present in mammalian hearts [3] .Anderson’s unifying criterion is that the entire AV junction be connected to one ventricular chamber, which includes hearts with one absent AV connection [4].

About ten percent of children with CHD (congenital heart disease) have functionally univentricular hearts. [5, 6]. A New England registry reported the incidence of univentricular heart to be 54 cases per million live births [7]. Hypoplastic left heart syndrome (HLHS)alon is the most common form of univentricular heart [8].

Tricuspid atresia (TA), the second most common subtype of univentricular heart, is thought to occur less than once for every 10 000 live births, and was present in 2.9% and 1.4% of congenital heart disease autopsy and clinical series, respectively [9].

Tricuspid atresia can be classified according to the morphology of the valve, [10, 11] the radiographic appearance of pulmonary vascular markings, [12, 13] and the associated cardiac defects [14-17] , but usually are classified in a manner initially proposed by Kuhne [18] and later modified [19].

Double inlet left ventricle (DILV) comprises 1% of all congenital heart malformations [20]. In an autopsy series of 60 univentricular hearts that excluded mitral and tricuspid atresia, DILV was present in 78%, double inlet right ventricle in 5%, and single ventricle heterotaxy syndrome in 13% [21].

Double outlet right ventricle (DORV) accounts for less than 1% of all congenital heart defects. Its incidence is approximately 0.06 cases per 1,000 of live births [22]. The ventriculoarterial relationship is defined by the ventricle from which more than 50% of an overriding semilunar valve originates [23].

In the setting of univentricular heart of left ventricular morphology (with exception of TA), the majority of patients will have discordant ventriculoarterial connections, with the aorta arising from the rudimentary outlet chamber of right ventricular morphology, and the pulmonary artery arising from the dominant ventricular mass of left ventricular morphology [23].

Pulmonary outflow tract obstruction frequently occurs with a univentricular heart. This can occur with either concordant or discordant ventricular-arterial connections [23].

The aim of this study is to determine the echocardiographic morphological classification of patients with univentricular heart disease and the subclassification of each type in Sulaimani governorate/ Kurdistan region/Iraq.

Table 1. Classification of Tricuspid atresia.

Type I	Normally related great arteries
	a. Intact ventricular septum with pulmonary atresia
	b. Small VSD and pulmonary stenosis
	c. Large VSD without pulmonary stenosis
Type II	Transposition of great arteries
	a. VSD with pulmonary atresia
	b. VSD with pulmonary stenosis
	c. VSD without pulmonary stenosis
Type III	Transposition or malposition of the great arteries. Associated complex lesions, that is truncus arteriosus, atrioventricular septal defect.

PATIENTS AND METHOD

This is a retrospective study conducted in pediatric cardiology unit /Sulaimani Pediatric teaching hospital from September 2009 to January 2014. All children with confirmed diagnosis of single ventricle anatomy were included in the study, hypoplastic left heart syndrome have been excluded from the study.

The source of information was the medical and echocardiography records from pediatric cardiology unit of Sulaimani city teaching hospital. For each patient study the following measures were recorded:

Two dimensional and Doppler (spectral and color) echocardiography examinations were obtained for each patient using a commercial instrument with 3V2C and 7V3C MHZ transducers (adjusted according to patient chest wall thickness) Acuson Cypress, USA made, supplied by Siemens Company. The echo measurements were recorded according to the standards recommended by the American society of Echocardiography and defined accordingly which usually allows clear demonstration of all the intracardiac anatomy.

All patients were studied by the same operator (pediatric cardiologist)in supine position and sedation depending on the state of the patient. Various echocardiographic views were used from parasternal, apical, and subcostal windows. Examinations usually started with a subcostal view in an attempt to demonstrate situs and interatrial septum and interventricular septum, chambers and relation to great artries and atrioventricular valves.

Atrioventricular valve morphology was then studied in a view equivalent to the apical four-chamber view, with the transducer placed at the cardiac apex. The "four-chamber" view was most useful for defining the number of atrioventricular valves, the papillary muscle relationships and the relationships of atrioventricular valves to inferior rudimentary chambers. Both the short-axis plane near the level of the great arteries or near the atrioventricular junction and subcostal planes were used to define the position and size of the rudimentary chamber.

While performing and reviewing these studies, we attempted not to diagnose univentricular hearts only , but also to subclassify the univentricular hearts according to atrioventricular connections, Ventricular septal defect(VSD), great artery malpositions (transposition, double outlet), and anatomic aspects of main-chamber morphology.

Studies also evaluated for specific associated abnormalities, including stenosis or atresia of semilunar valves, right and left isomerism, presence of Patent ductus artriosus(PDA), and total anomalous pulmonary venous return(TAPVR).

We depend on modified Kuhne [19] classification of Tricuspid Atresia.

According to Van Praagh [3] classification of DILV based on the great artery relationships, we did classification of DILV. Also according to Moss and Adam's textbook classification of DORV and other studies [24-26] we did DORV classification, and we depend on Van Praagh [2] classification of common AV valve univentricular hearts. Aortic atresia and HLHS were not included in the study as most of deaths happen in few days of life. All classifications were anatomical classifications.

Each returned questionnaire was given an identity number. Prior to data entry and analysis, the questions of study were coded. The data was entered into a Microsoft Excel Spreadsheet. After data cleaning, the data was transported into SPSS (Statistical Package for the Social Sciences – version 17) package software program for statistical analysis. Descriptive statistics (numbers and percentages) were calculated for all variables and its relation to each others or related findings .

Limitations and difficulties was related to variety of defects and their details but patients all were seen by pediatric cardiologist operator and confirmed by other coming as team for helping.

RESULTS

A combination of observations was used to diagnose univentricular heart. These included the number of atrioventricular valves, the position of the atrioventricular valves within the main chamber and separated from the rudimentary chamber, and failure to image a definable inlet septum between the atrioventricular valves or near the apex of the heart.

Total number of 100 patients, from neonatal period to 18 years (with one case 24 week gestational age fetus) at the time of study were diagnosed by two-dimensional echocardiography as univentricular heart patients. 55 cases were male, and 45 cases were female. 20 patients underwent angiocardiography and operation. 15 cases were deceased.

Among 100 cases, 73 cases (73%) had univentricular heart of left ventricular morphology, 24 cases (24%) had univentricular heart of right ventricular morphology, and 3 cases (3%) had univentricular heart of indeterminate type, as shown in table (1).

Among 100 cases, 50 cases (50%) were Tricuspid Atresia, 22 cases (22%) were DILV, 17 cases (17%) were DORV, 10 cases (10%) were common AV valve, and one case wasDouble inlet right ventricle(DIRV) (1%), as shown in table (2).

In this study from the sum of 100 cases, 85 cases (83%) were lived, 15 cases (15%) were deceased within four years of follow up (9 of them with TA, 3 cases with DILV, and 3 cases with common AV valve).

Subclassification of each type

a.Classification of patients with Tricuspid Atresia:

Type I patients were 40 cases (80%), type II patients were 10 cases (20%), type III has not been identified in our cases (Table 3).

Another case is 24 week gestational age diagnosed by fetal echocardiography as type 1-a. with a history of another sibling had the same condition, who passed away in infancy.

b. Classification of patients with DILV:

- Type 1 (normally related GA, Holmes heart): 1 case (4.54%)
- Type 2 (right-anterior aorta): 9 cases (40.9%)
- Type 3 (left-anterior aorta): 12 cases (54.54%)
- Type 4 (left-posterior aorta): none. As shown in figure (1).

3 patients had dextrocardia, and one patient with TAPVR.

c. DIRV:

One patient with this rare type, associated with pulmonary stenosis and dextrocardia.

d. DORV:

- Tetralogy type: 10 cases (58.8%).which means single anatomically RV with subvalvular PS and anterior alignemnt (TOF morphology)
- Transposition type: 5 cases (29.4%).
- VSD type: 2 cases (11.76%).

e. Common AV valve:

- Type A: morphologic LV 2 cases (20%)
- Type B: morphologic RV 5 cases (50%)
- Type D: ventricle with undifferentiated myocardium 3 cases (30%).

Right isomerisms with asplenia were present in 3 cases (30%). Left isomerisms with polysplenia were present in one case (10%). Situs inversus in one case (10%). 3 patients had transposition of great arteries (TGA), 3 patients had TAPVR, and one patient had dextrocardia.

Isomerism, 8 cases had isomerism, 5 cases with right isomerism (3 cases were male and 2 cases were female). 3 cases had left isomerism, two cases were female (with polysplenia), and one case was male.

Associated anomalies (Table 4):

- Among 100 patients with univentricular heart disease, 39 cases (39%) had transposition of great arteries. 32 cases (82.1%) with univentricular heart of left ventricular morphology, and 7 cases (17.9%) with univentricular heart of right ventricular morphology.
- Atrial situs: 91 cases (91%) showed atrial situs solitus, 8 cases (8%) had situs ambiguous, and one case (1%) had situs inversus.
- Dextrocardia : 8 cases (8%)
- Pulmonary Valve: 54 cases (54%) had pulmonary valve stenosis and atresia.
- ASD: 44 cases (44%)
- VSD: 50 cases (50%)
- Patent ductus arteriosus: 19 cases (19%).
- Pulmonary hypertension: 9 cases (9%) one of them became Eisenmenger syndrome on follow up. pulmonary banding had been done for 2 of the patients.
- TAPVR: 5 cases.
- PAPVR (partial anomalous pulmonary venous return): 1 case.

Table 1. classification according to the site of dominant ventricle.

Ventricular morphology	Number
Left ventricular morphology	73
Right ventricular morphology	24
Undetrmined ventricle	3

Table 2. Classification according to the main types of univentricular hearts.

Types of univentricular hearts	Frequency (%)
TA	50
DILV	22
DORV	17
AVSD	10
DIRV	1

Table 3. Classification of Tricuspid Atresia.

Tricuspid atresia	Subtype	No. of cases	%
I	Normally related great arteries	40	80%
	a. intact ventricular septum with pulmonary atresia	12	
	b. small VSD and pulmonary stenosis	23	
	c. large VSD without pulmonary stenosis	5	
II	Transposition of great arteries	10	20%
	a. VSD with pulmonary atresia	2	
	b. VSD with pulmonary stenosis	3	
	c. VSD without pulmonary stenosis	5	
III	Transposition or malposition of the great arteries. Associated complex lesions, that is truncus arteriosus, atrioventricular septal defect.	0	0

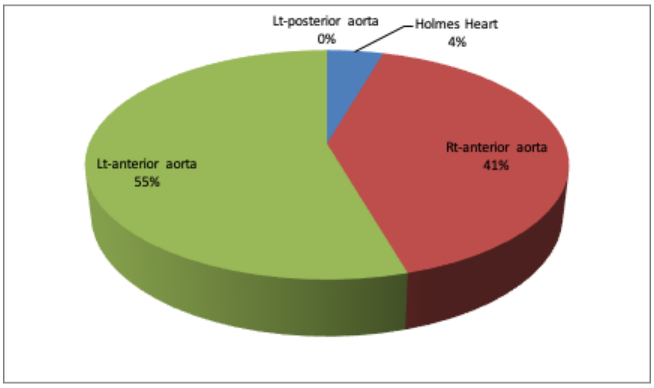

Figure 1. DILV subtypes.

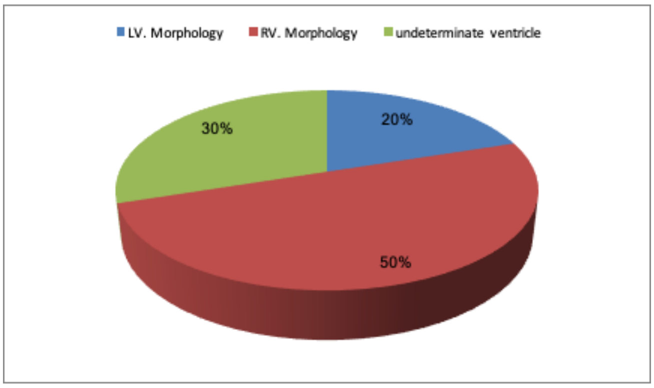

Figure 2. Common AV valve subtypes.

Table 4. Associated anomalies in univentricular heart patients.

Associated lesions		No.(%)
Position of great vessels		
Normal	LV morphology	44(44)
	RV morphology	17(17)
TGA	LV morphology	32(32)
	RV morphology	7(7)
Situs solitus		91(91)
Dextrocardia		8(8)
Dextrocardia with situs inversus		1(1)
AV association :		
concordant		76(76)
discordant		24(24)
Interatrial communication		44(44)
Right isomerism		5(5)
Left isomerism		3(3)
Interventricular communication		50(50)
Pulmonary stenosis	LV morphology	27(27)
	RT morphology	5(2)
	Indeterminate type	2(2)
Patent ductus arteriosus		19(19)
TAPVR		5(5)
PAPVR		1(1)

DISCUSSION

This study is the first study done in Iraq up to our knowledge. The results of our study suggest that two-dimensional echocardiography is very useful for noninvasive diagnosis of univentricular heart. In most patients, it can accurately evaluate atrioventricular connections as well as the morphology of the main chamber and the outlet chamber. Accurate diagnosis of univentricular heart by two-dimensional echocardiography does aid in planning a hemodynamic study, and localization of outlet chambers aids in planning angiographic evaluation.

Our finding that the TA is the most common type of univentricular heart disease (with exclusion of HLHS) is consistent with other studies [9].

Regarding subtypes of TA, type I constitute 80% of the cases, which is the most common subtype of TA, and type II 20% of the cases, and this is consistent with Moss and Adam's textbook and other studies [27, 28] in which it was mentioned that type I occurs in 70-80% , and type II occurs in 12-25% of cases with TA.

In patients with DILV, 94% of them have discordant VA connection (TGA), and this is nearly consistent with a study done by Shiraishi H and Silverman NH and Maurizio Bevilacqua et al. [29, 30] in which their study shown that 86% of patients with DILV had TGA, and the 3rd type (left-anterior aorta) is the most common type.

DIRV found only in one patient, while it was found in two patients in the series reviewed by Van Praagh et al. [3, 21].

Regarding subtypes of DORV, TOF(tetralogy of Fallot) type is the most common type (59%), followed by Transposition type (29%), and the less common type is the VSD type (12%). This is nearly consistent with other studies done about DORV [31, 32] in which TOF type seen in approximately 40% of cases, transposition type in 20% of cases, and VSD type in 15% of cases.

In patients with common AV valve, 30% of them have asplenia (our sample size is small). In the series of Van Praagh et al. [3, 21] 40% of the cases with common AV valve have asplenia.

In patients with heterotaxy syndrome (isomerism), the right isomerism with asplenia (62%) were more common than left isomerism with polysplenia (37.5%). In comparison to Van Praagh et al. review [33] in which 59% of patients with heterotaxia were asplenia, and 39% were polysplenia, both studies revealed that the right isomerism was more common.

The right isomerism was more common in male patients, while left isomerism was more common in female patients in our study. This is consistent with what was mentioned by Satpathy M [34].

Associated anomalies: in our study, VSD is the most common associated anomaly, followed by ASD. With exclusion of TA and DORV, about 63% of cases (DILV and Common AV valve) have TGA; while in a study [35] 84% of the cases with DILV and Common AV valve have TGA.

In our study, one fetus had been found on fetal echocardiography having TA, and the mother had had a history of another child with TA who passed away during infancy. This will rise a concern about genetic markers that may be responsible for this defect (no specific genes has been found yet) [36].

Because of high prevalence cases of CHD and high drainage of patients from cities around, especially those with complex congenital heart diseases in which they need early intervention, we suggest the importance of presence of pediatric cardiac hospital in Sulaimani city for taking care of these patients for doing cardiac catheterization and surgical palliative care with follow up.

In conclusions, our study proved that univentricular heart disease is not uncommon, the TA is the most common type of univentricular heart (with exclusion of HLHS). Right isomerism is more common than left isomerism. Right isomerism is more common in male patients. VSD & ASD are the most common associated anomalies. TA with normally related great arteries is the most common type of TA. Transposed great arteries are the most common subtype of DILV. TOF is the most common subtype of DORV in our study.

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