



Research Article

**UNOPERATED SINGLE VENTRICULAR PATIENT WITH PROSTHETIC VALVE:
A RARE CASE-REPORT**

Cetin Sanlialp Sara

Department of Cardiology, Servergazi State Hospital, Denizli, Turkey

ARTICLE INFO

Article History:

Received 15th February, 2019

Received in revised form 7th

March, 2019

Accepted 13th April, 2019

Published online 28th May, 2019

ABSTRACT

Univentricular heart (UVH) is a complex and congenital cardiac malformation that both atria are connected to a functional single ventricle (1). The prognosis is poor when this malformation is not operated (2). Although rare cases have been reported in literatures, surviving into adulthood without surgery is mostly difficult. This article is about an adult patient with single ventricle who was not operated, but the patient had a prosthetic valve and the coexistence of these two situations make the case very interesting.

Key words:

adult, congenital, long-term survival, single ventricle

Copyright©2019 Cetin Sanlialp Sara. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

INTRODUCTION

Responsible for 1--2% of all congenital heart defects (3,4). UVH has various physiological and functional defects (5). The patients with UVH survive into adulthood by modern surgical technique, cardio protection and perioperative management in today (6). But unoperated UVH causes mostly death in infants (7) and it is almost unusual to survive into late adulthood (8). In this case, we discussed an asymptomatic adult the patient had unoperated single ventricle who had prosthetic valve.

Case Report

46-year-old asymptomatic male patient admitted to the outpatient clinic for general control and warfarin dose adjustment. The patient have unoperated single venticle and when this malformation noticed, he was eighth years. Although the operation was suggested, his family did not accept due to high risk for complications and mortality. In the following years, the patient was operated because of serious mitrale regurgitation. He only took warfarin treatment due to mechanic mitrale valve. Physical and mental development of the patient was normal. He had 178 cm height and 75 kg weight. The patient's fingers were mild clubbing but the general appearance of the patient was not cyanosis. His blood pressure was measured as 120/60 and pulse 72 / min. His oxygen saturation was 95% with pulsi oximeter and his breath rate was normal. 3/6 systolic ejection murmur and metallic valve sound were detected in the apical region in oscultation.

His electrocardiyogram was sinus rhtym and right axis. His cardiothropic index was compatible with cardiomegaly in telecardiography. In transthoracic echocardiography, apical four-chamber examination; two separate atrioventricular valves opened to a single ventricular cavity and there was a bulboventricular foramen. Also it was observed that rudimenter ventricular cavity was superior and front position which is called outlet chamber. Other features on echocardiography were repectively functional prosthetic mechanic valve, mild pulmoner stenosis and moderate pulmoner artery pressure (42 mmhg).The ejection fraction was measured as 47% by Simpson method. The patient was diagnosed with double inlet morphological left ventricle with current echocardiographic findings. According to the valve operation epicrasis, the patient had normal related with great arteries vessels. Low dose ACEI/ARB was added to warfarine for optimum medical treatment. Infective endocarditis prophylaxis and cardiac catheterization were recommended to patient.

DISCUSSION

A single ventricle is characterized as lacking two well-developed ventricles (9). In today the new term that is labeled functional univentricular heart has been created (10). Fuctional UVH can be defined that only one ventricle provides systemic and pulmoner circulation of heart (11). Single ventricle can be classified according to great arterial positions, pulmoner atresia or stenosis, AV connection and ventricular morphology (12). If single venticle has biventricular AV connection, it is designated doublet inlet, if it has no AV connection, it can be mentioned undetermine ventricle (13). The ventricle morpology can be classified according to the shapes of

*Corresponding author: **Cetin Sanlialp Sara**

Department of Cardiology, Servergazi State Hospital, Denizli, Turkey

ventricle, rudimentary ventricular position and the presence of infundibulum. If the ventricle wall is straight and it has anterosuperior placement rudimentary ventricle and finally single ventricle does not include infundibulum, the morphology of ventricle is left. Unlike if the ventricle has trabecular structure and does not include infundibulum or posteroinferior placement rudimentary ventricle, it is right ventricular morphology. When the ventricle does not provide these morphologic properties, it is called undetermined ventricle (14). The left ventricular type is most common in patients (60%-66%). The incidence of other types are 10%-24% for right ventricular morphology and <10% for indeterminate type. (15)

Figure. Echocardiographic apical 4-chamber imaging with single ventricle and prosthetic valve



The long term survival and clinical presentation of patients depend on pulmonary blood flow, pulmonary vascular resistance, aortic flow, ventricular morphology, functions of ventricle and the presence / absence of AV valves. In presence of moderate pulmonary stenosis (if Qp is nearly normal), mild or no heart failure and mild systemic desaturation; the long-term survival may be possible because of providing the balance of adequate oxygenation (pulmonary circulation) and volume load of the ventricle (systemic circulation). On the other hand there are no reports of long term survival with right and undetermined ventricle type. Also the presence of separated AV valves with no moderate or severe regurgitation and the absence of critical subaortic obstruction may effect the long-term survival (16). In this case, the patient had properties that affect survival positively such as moderate pulmonary stenosis (SPAB: 42 mmHg), doublet inlet left ventricle type and nearly normal cardiac systolic function. Also he had no moderate or severe valve regurgitation except prosthetic valve. But his ventricle can be effect overload volume before mitral valve replacement even if he was asymptomatic before surgery. It can be only explained in this way; he had moderate or severe regurgitation and then the regurgitation caused over volume load. Finally his ventricular systolic function mild decreased due to over volume load. Nevertheless he had long-term survival because of well balanced two circulation systems.

Life expectations of patients without surgery is 4-14 years (8). The oldest single ventricular adult patient in the literature belongs to Goldenberg (17). There are very few adult patients (50-60 years) with single ventricle in the world have been reported (16). Ammash *et al.* (1996) specified that patients that had well developed pulmonary circulation can reach into six

decades with good functional capacity (18). Four patients with 21, 23, 28 and 45 age years were reported exclude our patient yet in Turkey (19). Although surgery is recommended in childhood; atrial arrhythmia, venous congestion, protein-losing enteropathy, thromboembolism and heart failure may occur post operative period and long-term (16). The most common causes of death are arrhythmia, heart failure and sudden cardiac death (19). But the operation in early ages is associated with better exercise tolerance and quality life. In adults with front of fontan operation, the quality of life, physical function, mental health is lower (1). So older adult patients may be followed and treated conservatively because of high morbidity and mortality risks of operation (16).

We agree that the patient had long-term survival due to left morphologic type ventricle, well developed pulmonary vascular system and nearly normal systolic functions. However, we did not recommend him operation because he was asymptomatic and his exercise capacity was good. Also the operation was associated with increased mortality and complication risk. Moreover he might not benefit from surgery due to his adult age. So only we informed him about arrhythmic, neurological and hematologic problems in long term.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Paul Khairy, Nancy Poirier *et al.* Univentricular Heart. *Circulation*. 2007;115:800-812.
2. Kenan Demir, Hakan Akilli. A rare case of congenital heart disease in an unoperated adults patient single ventricle. *Selcuk Med. Journal*. 2012;28(2):128-129.
3. M Samanek. Children with congenital heart disease: probability of natural survival. *Pediatr Cardiol*. 1992;13:152-158.
4. M Samanek, M Voriskova. Congenital heart disease among 815,569 children born between 1980 and 1990 and their 15-year survival: a prospective Bohemian survival study. *Pediatr Cardiol*. 1999;20:411-417.
5. Ertugrul Gök, Okan Akan, Bülent Eren *et al.* The heart with single ventricle detected by exhumation and forensic autopsy: A case report. *Dicle Medical Journal*. 2015;42(4):522-524.
6. Sanaz Piran, Gruschen Veldtman *et al.* Heart failure and ventricular dysfunction in patients with single or systemic right ventricles. *Circulation*. 2002;105:1189-1194.
7. Oliver JM, Fdez-de-Soria R, Dominiguez FJ *et al.* Spontaneous long-term survival in single ventricle with pulmonary hypertension. *Am Heart J*. 1990;119:201-202.
8. Moodie DS, Ritter DG, Tajik AJ *et al.* Long-term follow-up in the unoperated univentricular heart. *Am J Cardiol*. 1984;53:1124-1128.
9. Jacobs ML, Mayer JE Jr. Congenital Heart Surgery nomenclature and database project: single ventricle. *Ann Thorac Surg*. 2000;69(4):197-204.
10. Anderson RH, Cook AC. Morphology of the functional univentricular heart. *Cardiol Young*. 2004;14(1):3-12.

11. GS Saylam, A Sarıoglu. Fonksiyonel tek ventriküllü hastalara yaklaşım ve tedavi seçenekleri. *GKD Cer Derg.* 1996;1:15-27.
12. Driscoll DJ, Schaff HV. Single ventricle. *Cardiology fundamentals, practise.* 2 nd ed. St Louis: Mosby-Year Book:1991.
13. Carla Frecsura and Gaetona Thiene. The new concept of univentricular heart. *Journal Frontiers in Pediatrics.* 2014;2:62.
14. Serkan Arıbal, Hakan Önder. Pediyatrik Konjenital Hastalıkları. *Trd Sem.* 2018;6:233-248.
15. JK Perloff. The clinical recognition of congenital heart disease. (4 th ed), WB Saunders, Philedelphia.1994.
16. Alfred Hager, Harald Kaemmer *et al.* Long-term survival of patients with univentricular herat not traeted surgically. *The Journal of Thoracic and Cardiovascular Surgery.* 2002;123(6):1214-1217.
17. HL Goldenberg *et al.* Prolonged survival (62 years) with single ventricle. *Am J Cardiol.*1983; 52:214-215.
18. NM Ammash, CA Warnes. Survival into adulthood of patients with unoperated single ventricle. *Am J Cardiol.* 1996;77(7):542-544.
19. Bilge Duran Karaduman, Hüseyin Bayram, Hacı Ahmet Kasapkara, Telat Keleş, Tahir Durmaz. Ameliyat edilmeden erişkin yaşa ulaşmış tek ventriküllü olgu. *Türk Kardiyol Dern Ars.* 2016;44(4):338-341.

How to cite this article:

Cetin Sanlıalp Sara (2019) 'Unoperated Single Ventricular Patient with Prosthetic Valve: A Rare Case-Report', *International Journal of Current Advanced Research*, 08(05), pp. 18617-18619. DOI: <http://dx.doi.org/10.24327/ijcar.2019.18617.3564>
