

Long-term survival of a patient with single atrium and single ventricular heart

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Introduction

Uni-ventricular heart is rare and accounts for 1-2% of all congenital cardiac malformations. Without corrective surgery, it has a fatal course in the neonatal period or in early infancy and survival into adult life is unusual [1]. Majority of patients with uni-ventricular heart have to undergo surgical procedures during infancy or childhood to ensure a balanced pulmonary and systemic blood flow. Although, for a long time, Fontan-type procedures have been performed as corrective surgical treatment, it is also associated with significant mortality and morbidity due to complications such as atrial arrhythmia, venous congestion, protein-losing enteropathy, thromboembolism and ventricular failure [2]. However, rarely some patients live almost a normal life without any surgical interventions due to a balanced native hemodynamic status. Only around 10 cases have been reported worldwide who survived into late adulthood; age up to 62 years in one and up to 50 years in the rest [3]. We did not find any reports from Sri Lanka of long survival of such patients.

Case report

A 25-year-old female patient with a single ventricle, common atrio-ventricular valve with moderate to severe valve regurgitation, severe pulmonary stenosis, left malposition aorta, with large ostium primum atrial septal defect (Single atrium) was admitted to our unit.

She was born without any complications. Antenatally mother didn't have any diseases. After 14 days of birth she had been admitted to hospital due to a lower respiratory tract infection and central cyanosis where the above cardiac diagnosis was made. At the age of 10 years she was evaluated with 2D echocardiogram, cardiac catheterization and angiogram with view to carrying out surgery. Due to severe atrio-ventricular valve regurgitation

and high normal pulmonary pressure, direct total Fontan surgery was contraindicated. Other corrective options such as bidirectional Glenn surgery with atrio-ventricular valve repair to arrest the regurgitation were also considered unhelpful. Since the oxygen saturation was around 85%, it medical follow-up was planned.

Her growth and development during infancy and childhood were documented normal. Although she was average in studies and managed daily activities without difficulty, she couldn't take part in sports due to reduced exercise tolerance. After leaving school, she started to work in a textile shop.

During the current admission, she complained of intermittent palpitation and dyspnea of over 3 months duration. Physical examination found breathlessness, moderate cyanosis and clubbing. Radial pulses were feeble with the rate of >120 beats/minute, blood pressure was 90/60 mm Hg with elevated jugular venous pressure. The apex of the heart was shifted inferiorly and laterally and there was a systolic thrill all over the precordium. A grade 4-5 systolic murmur was heard all over the precordium. Respiratory examination showed bi-basal inspiratory fine crepitations. Liver and spleen were not palpable.

Electrocardiography showed supraventricular tachycardia with a ventricular rate of 120 beats/minute and was successfully treated with IV amiodarone. Chest radiography showed cardiomegaly. Transcutaneous oxygen saturation was 86%. Haemoglobin level was 17.6 g/dl and haematocrit was 54%. All the other investigations were within normal limits.

Transthoracic 2D echocardiography showed previously documented cardiac abnormalities; single ventricle, large ostium primum atrial septal defect and atrio-ventricular valve gradient >100 mm Hg with no coarctation of the aorta (Figure 1). Cardiac catheterization

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was considered not essential. She was discharged with oral amiodarone and remained clinically and haemodynamically stable on follow up after two weeks.

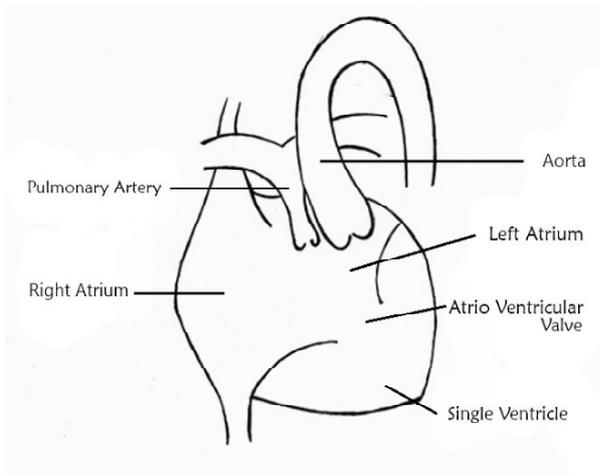


Figure 1. Diagrammatic representation of the heart with common AV valve, single ventricle and large ostium primum atrial septal defect.

Discussion

Uni-ventricular heart is a rare congenital cardiac anomaly and is categorized based on the ventricular anatomy; left ventricular type (60-66%), right ventricular type (10-24%) and intermediate type (<10%) [4]. The great vessel connections are variable and many other anomalies may coexist. The clinical presentation and long-term outlook depends on the presence or absence of an

obstruction to the pulmonary blood flow, pulmonary vascular resistance, morphology and function of the ventricle and atrio-ventricular valve and the degree of obstruction to aortic flow.

Review of patients with long survival demonstrate that patients with uni-ventricular heart and well-balanced pulmonary perfusion might survive into late adulthood with good quality of life and functional capacity, without major symptoms or depression of cardiac function. The most promising survival is seen with left ventricular type morphology, transposition of the great arteries without systemic outflow obstruction, an adequately functioning atrio-ventricular valve, and a moderate pulmonary outflow obstruction [5]. The reason behind the long term survival in our patient could be due to similar cardiac abnormalities that had been previously described in long term survivors.

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