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Brief Report

A 74-year-old unoperated univentricular heart: the oldest reported survival

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Abstract Univentricular heart is a rare congenital heart malformation. Nowadays prognosis is considered to be strictly linked to surgical intervention, and survival into late adulthood is unusual.

In some patients native haemodynamic circulation balances pulmonary and systemic blood flow, allowing long-term survival without the need for surgery.

We report the case of a 74-year-old man with a univentricular heart in natural history, and we discuss the factors that might contribute to his extraordinary long-term survival.

Keywords: Grown-up CHD; univentricular heart; natural history

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Case report

A 74-year-old man was admitted to our institution for cardiac monitoring after orthopaedic intervention for a medial fracture of the femur secondary to an accidental fall. He reported a history of cyanosis noted a few months after birth. Although Congenital Heart Disease (CHD) was suspected, his underlying anatomy was not comprehensively evaluated until cardiac catheterisation at 18 years. At that time he was diagnosed with dextrocardia, a univentricular heart, transposition of the great arteries, and pulmonary outflow obstruction. Surgical intervention was not offered. His growth and development during infancy and childhood were considered normal, but he reported a history of therapeutic phlebotomies that were performed during the 1960s and 1970s when his haematocrit exceeded 65%.

When he was 58 years old, he experienced an episode of atrial fibrillation successfully treated with electric cardioversion. Oral anticoagulant therapy was not started because of polyglobulia.

Over the years he gradually presented decreased exercise tolerance.

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When he was 69 years old, he experienced one episode of heart failure with dyspnoea, worsening of cyanosis, and recurrence of atrial fibrillation. He was treated with diuretics and β-blockers and was begun on dicumarol therapy. Thereafter he was clinically stable, with NYHA grade II.

His physical examination on admission showed O_2 saturation 75–80%, digital clubbing, and a 3/6 L right-sided parasternal systolic murmur. The liver and spleen were normal; peripheral oedema was absent. The lungs were clear. Blood pressure was 110/70 mmHg.

His electrocardiogram showed atrial flutter with a ventricular rate of 60–80 beats/minute and inferolateral Q waves, consistent with a dextrocardia pattern. Chest radiography showed enlarged pulmonary arteries without signs of congestion. Laboratory tests after orthopaedic surgery showed haemoglobin 13.9 g/dl, haematocrit 43.3%, and mean corpuscular haemoglobin 25.9 pg.

Echocardiography demonstrated atrial situs solitus, dextrocardia, D-looped ventricles, double inlet left ventricle – two atrioventricular valves connected to a single morphological left ventricle with normal function: ejection fraction 67% and end-diastolic volume 85 ml/mq – a large atrial septal defect ostium secundum, non-restrictive bulboventricular foramen, L-transposed arteries – anterior

left-sided aorta arising from a small right ventricular outlet chamber without obstruction and a posterior right-sided pulmonary artery connected to the morphological left ventricle – and moderate subvalvular and valvular pulmonary stenosis with a maximum gradient of about 60 mmHg. A mild-to-moderate regurgitation of the right-sided atrioventricular valve was present.

During hospitalisation, he underwent a rehabilitation programme without cardiac events.

Currently, 1 year later, he is stable.

Discussion

Univentricular heart is a rare congenital anomaly that accounts for about 1% of all CHDs at birth.[1] It is defined as the presence of a single ventricular chamber or two ventricular chambers in which one is severely hypoplastic. Associated anomalies are pulmonary stenosis or atresia, subaortic stenosis, coarctation of aorta, and mitral or tricuspid atresia. It is usually associated with transposed great arteries, whereas Holmes' heart is a rare variety in which the great arteries are normally related.[2]

In unoperated univentricular heart patients the survival rate in the 1st year of life is ~30%[3] and the median survival is 14 years.[4] Survival into late adulthood without surgical repair is rarely reported.

The clinical presentation and long-term outcome depend on the presence of obstruction in the pulmonary blood flow and pulmonary vascular resistance, morphology, and function of the ventricle, obstruction to aortic flow, and morphology and function of the atrioventricular valves.[5]

A moderate pulmonary stenosis is crucial for haemodynamic balance in this CHD: on one hand it limits the pulmonary blood flow, avoiding pulmonary arteriolar disease, and on the other it causes systemic desaturation. Long-term survival is possible with adequate oxygenation and a balanced ventricular load. Pulmonary stenosis limits the left to right shunt that leads to an increased pulmonary flow and ventricular overload, finally protecting from heart failure.

Morphology and function of the single ventricle are also critical for the clinical course in univentricular hearts: there are, in fact, no reports of long-term survival with right morphology or a indeterminate-type single ventricle. This is probably due to the intrinsic capacity of the left ventricle to deal with the systemic resistance.

Survival is positively influenced by characteristics of atrioventricular valves: competent and separated valves are linked to a better outcome.[5] Incompetent atrioventricular valves impose a volume overload on the ventricle.

We reviewed the literature on unoperated univentricular hearts and found that atrial arrhythmias, such as typical atrial flutter and atrial fibrillation, are the most frequent complications among patients who survive longer, but to our knowledge there is no report on the mean age at onset[6] (Table 1). We found 13 patients:[5,6,13,14] three presented with a history of paroxysmal atrial fibrillation/flutter that was medically controlled, two had permanent atrial fibrillation, and one underwent multiple electrical cardioversions and an ablation procedure for intra-atrial re-entry tachycardia, whereas our patient had atrial flutter.

Table 1. Demographic and clinical data and cardiac anatomy of patients with univentricular heart in natural history surviving beyond the age of 50 years.

References	Age/Sex	Type	Arrhythmias	Clinical condition
Goldberg et al[7]	62/Male	DILV, TGA, PS	–	Died
Oliver et al[8]	61/Female	DILV, MGA, PHT	–	Mild symptoms
Habeck et al[9]	59/Male	DILV, TGA, PHT	Atrial fibrillation	Died
Koito et al[10]	57/Male	DILV, TGA, PHT	–	Alive
Warner et al[11]	61/Female	DILV, TGA, PS, endocarditis	–	Surgical treatment
Ammash and Warnes[12]	60/Male	DILV, TGA, PS	–	Died
Ammash and Warnes[12]	60/Male	DILV, TGA, PS	Paroxysmal atrial fibrillation	Mild symptoms
Ammash and Warnes[12]	66/Male	DILV, TGA, PS	Paroxysmal atrial flutter	Fairly good
Gabbarini[2]	61/Male	DILV, MGA, PHT	Paroxysmal atrial fibrillation	Fairly good
Hager et al[5]	62/Female	DILV, TGA, PS	Atrial fibrillation	Mild symptoms
Restaino et al[13]	57/Female	DILV, TGA, PS	–	Mild symptoms
Terada et al[14]	60/Female	DILV, TGA, PS, endocarditis	–	Died
Dhillon et al[6]	62/Male	DILV, TGA, PS	Intra-atrial reentry tachycardia	Died
Present case	74/Male	DILV, TGA, PS	Atrial flutter	Mild symptoms

DILV = double inlet left ventricle; MGA = ventriculoarterial concordance with a left/right position of the aorta; PHT = pulmonary hypertension; PS = pulmonary stenosis; TGA = transposition of the great arteries with a left/right position of the aorta

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Dhillon et al[6] hypothesised that a progressive atrial myopathy, secondary to uncorrected CHD, leads to atrial fibrosis and may contribute to the genesis of arrhythmias in these patients.

The perspective of survival is greater in patients with the following associations:[5] single ventricle with left ventricular morphology; transposition of the great vessels without flow obstruction; atrioventricular valve working properly; and moderate obstruction to the pulmonary flow, which allows enough pulmonary flow to prevent severe cyanosis and avoid ventricular overload.

To our knowledge the literature reports only 13 patients with a univentricular heart in natural history who survived beyond the age of 50 years; all of them had left ventricular morphology. Compared with those patients, our patient had the same ventricular morphology but he is to date the oldest (maximal age, respectively, 62 versus 74 years).

The "ideal" anatomy seems characterised by left ventricular morphology, good ventricular function, and "well-balanced" circulation, with some degree of pulmonary stenosis that avoids at the same time excessive pulmonary blood flow and severe cyanosis. These anatomic characteristics can allow patients to survive with mild-to-moderate symptomatology and with a good quality of life. Cardiac arrhythmias are the most frequent complications among patients who survive longer. The patient presented here had these anatomic features, enabling him to reach the age of 74 years without surgical intervention.

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Conflicts of Interest

None.

References

1. Steinberg EH, Dantzker DR. Single ventricle with severe pulmonary hypertension: natural survival into the third decade of life. Am Heart J 1993; 125: 1451–1453.
2. Gabbarini F. A 61-year-old man with Holmes' heart. Lancet 1999; 353: 646.
3. Samanek M. Children with congenital heart disease: probability of natural survival. Pediatr Cardiol 1992; 13: 152–158.
4. Moodie DS, Ritter DG, Tajik AJ, O'Fallon WM. Long-term follow-up in the unoperated univentricular heart. Am J Cardiol 1984; 53: 1124–1128.
5. Hager A, Kaemmerer H, Eicken A, Fratz S, Hess J. Long-term survival of patients with univentricular heart not treated surgically. J Thorac Cardiovasc Surg 2002; 123: 1214–1217.
6. Dhillon PS, Li A, Gonna H, Waed DE. Intra-isthmus reentry associated with uncorrected double inlet left ventricle and transposition of the great arteries. Congenit Heart Dis 2013; 8: E56–E60.
7. Goldberg HL, Sniderman K, Devereux RB, Levin A. Prolonged survival (62 years) with single ventricle. Am J Cardiol 1983; 52: 214–215.
8. Oliver JM, Fdez-de-Soria R, Dominguez FJ, Ramos F, Calvo L, Ros J. Spontaneous long-term survival in single ventricle with pulmonary hypertension. Am Heart J 1990; 119: 201–202.
9. Habeck JO, Reinhardt G, Findeisen V. A case of double inlet left ventricle in a 59-year-old-man. Int J Cardiol 1991; 30: 119–120.
10. Koito H, Ohkubo N, Suzuki J, Iwasaka T, Inada M. Prolonged survival in a patient with a single ventricle without pulmonary stenosis. Chest 1994; 106: 971–972.
11. Warner KG, Payne DD, Fulton DR, et al. Excision of a pulmonary valve abscess in a 61-year-old woman with single ventricle. Ann Thorac Surg 1996; 61: 213–215.
12. Ammash NM, Warnes CA. Survival into adulthood of patients with unoperated single ventricle. Am J Cardiol 1996; 77: 542–544.
13. Restaino G, Dirksen MS, de Ross A. Long-term survival in a case of unoperated single ventricle. Int J Cardiovasc Imaging 1994; 20: 221–225.
14. Terada M, Watanabe H, Iino K, Kakizaki M, Takahashi M, Ito H. Adequate pulmonary stenosis allowed long-term survival in a patient with unoperated single ventricle. J Am Coll Cardiol 2012:59(13): e25.