Common atrium with single ventricle in a newborn: A case report

Irene Eseohe Akhigbe and Abubakarr Bailor Bah

Ola During Children's Hospital, University of Sierra Leone Teaching Hospital Complex, Freetown, Sierra Leone

Address for correspondence:
Dr Irene Akhigbe
Ola During Children's Hospital
University of Sierra Leone Teaching Hospital Complex
21 Percival Street
Freetown
Sierra Leone

Email:
drireney@yahoo.com

Irene Akhigbe ID: Not available yet
DOI: https://www.journals.ac.za/SAHJ/article/view/6822

ABSTRACT

The presence of a common atrium, a common atrio-ventricular valve in combination with a single ventricle, occurs very rarely, accounting for 1% - 2% of all congenital heart malformations.
We report a rare case of a 21-day old male neonate diagnosed with a common atrium, single ventricle, a common atrioventricular valve and a patent ductus arteriosus. High index of suspicion and early diagnosis is vital for appropriate management and timely surgical interventions as required. SA Heart® 2025;22:122-123

INTRODUCTION

Common atrium (CA) is defined as complete or near complete absence of the interatrial septum, and has clinical and haemodynamic similarities with large-size atrial septal defect of fossa ovalis type.[1-3] Common atrium is very rarely seen in patients with single ventricle (SV) accounting for 1% - 2% of all congenital heart malformations.[4,5] In patients with common atrium and SV, there is mixing of arterial and venous blood in the common cardiac chamber which causes severe cyanosis and hypoxia in these patients.[5,6]

CASE PRESENTATION

We report the case of a 21-day-old male newborn of non-consanguineous parents; that was referred to our facility with complaints of central cyanosis and feeding difficulties observed since birth, and he subsequently developed cough and dyspnoea by the 14th day of life. He was delivered via spontaneous vaginal delivery at 38 weeks of gestation, and had good extrauterine transition with a birth weight of 3 300 grams. His mother reported a positive history of febrile illness without rashes during early pregnancy, but there was no exposure to ionising radiation, no history suggestive of gestational diabetes, and she received only prescribed medications.

On examination he was cyanosed, not febrile, with no dysmorphic features or external malformations. Cardiovascular system examination revealed bounding peripheral arterial pulses, with a capillary refill time of <3 seconds and oxygen saturation of 77% in room air. The auscultatory findings included tachycardia, first and second heart sounds with a grade III/VI continuous murmur loudest in the second intercostals space, left upper parasternal border. He was dyspnoiec and tachypnoiec on respiratory examination with vesicular breath sounds. His abdominal examination demonstrated a palpable liver 4cm below the right costal margin that was soft, tender with a smooth surface.

An initial working diagnosis of a complex cyanotic congenital heart disease with patent ductus arteriosus in heart failure was considered. Chest radiography showed cardiomegaly with a cardiothoracic ratio of 0.69, increased intrapulmonary vascular markings and widened cardiac pedicle (Figure 1).

Echocardiogram revealed a patent ductus arteriosus, with a common atrium, common atrio-ventricular valve and a single ventricle of left ventricular morphology with a normal aorta and pulmonary artery arising from the single ventricle (Figure 2).

Patient was treated conservatively with furosemide, spironolactone, and captopril. His cardiac symptoms improved and was discharged on same care plan to the cardiology clinic. He remained clinically and haemodynamically stable on follow up visits, and awaits surgical evaluation due to financial constraints.

DISCUSSION

The single ventricle or univentricular heart occurs in ~5 in 100 000 newborns.[7] In most cases, both atria empty into a

FIGURE 1: Chest radiography showing cardiomegaly, increased intrapulmonary vascular markings and widened cardiac pedicle.

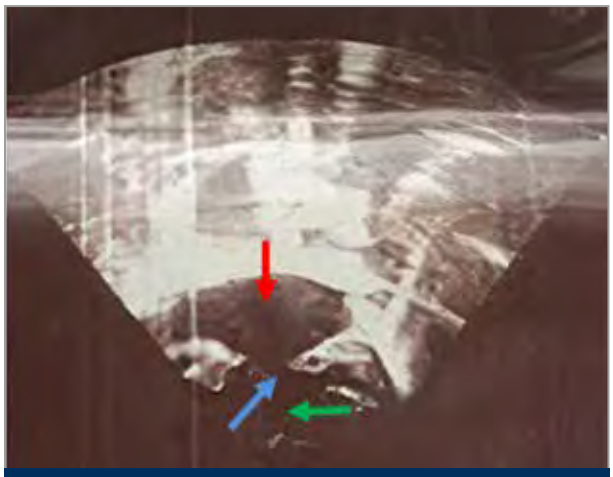

FIGURE 2: Transthoracic echocardiogram showing common atrium (red arrow), single ventricle (green arrow) and a common atrioventricular valve (blue arrow).

common ventricle through a separate atrioventricular valve. The presence of a common atrium, a common atrioventricular valve in combination with a single ventricle as in our case, occurs very rarely, accounting for 1% - 2% of all congenital heart malformations and was first described in 1842.[4,8]

The clinical presentation and long-term outlook depends on the presence or absence of obstruction to pulmonary blood flow, pulmonary vascular resistance, morphology and function of the ventricle and atrioventricular valve, and the degree of obstruction to aortic flow.[9] The median age of survival in a patient with right ventricle morphology is 4 years, while that of a left ventricle morphology is 14 years.[5,10,11]

Review of patients with long survival demonstrate that patients with univentricular 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.[12]

Patients however having decreased pulmonary blood flow would need a modified Blalock-Taussig shunt while those with increased blood flow may require a pulmonary artery banding procedure. This is usually followed up by a bidirectional Glenn operation as the second-stage after 3 - 6 months and a subsequent Fontan procedure at 2 - 3 years.[10] The surgical mortality of each case depends on the complexity of the lesion and the type of surgery planned at each stage of presentation and may range from 5% - 10% at each stage.[10]

CONCLUSION

We presented a case of a common atrium, a single ventricle with a common atrio-ventricular valve and patent ductus arteriosus, in a neonate. High index of suspicion and early diagnosis is vital for appropriate management and timely surgical interventions as required.

Conflict of interest: none declared.

REFERENCES

1. Levy MJ, Solomon J, Vidne BA. Correction of single and common atrium, with reference to simplified terminology. Chest. 1974;66(4):444-445.
2. Zhang Y, Yang ZG, Yang MX. Common atrium and the associated malformations: Evaluation by low-dose dual source computed tomography. Medicine®. 2018;97(46):e12983.
3. Bun SS, Squara F, Scarlatti D. Atrial fibrillation ablation in a single atrium with inferior vena cava interruption. Ann Noninvasive Electrocardiol. 2023; 28(4):e13057.
4. Drouin E, Marechaux S, Hautecoeur P. Univentricular heart with common atria. First historical cases. International Journal of cardiology congenital heart defects. 2021;6(9):100267.
5. Munoz-Armas S, Gorrin JR, Anselmi G. Single atrium: Embryologic, anatomic, electrocardiographic and other diagnostic features. Am J Cardiol. 1968; 21:639-52.
6. Kinare SG, Sivaraman A, Deshpande J. Single ventricle (morphologic study of 21 cases). Indian Heart J. 1989;41(5):301-306.
7. Frescura C, Thiere G. The new concept of univentricular heart. Front Paediatr. 2014;2:62.
8. Thore JÉ. A Simple Heart. Bulletins of the Anatomical Society of Paris. 1842;145.
9. Lohithalingam P, Lakmini KMS, Weerasinghe S, Pereira T, Premaratna R. Long-term survival of a patient with single atrium and single ventricular heart. Ceylon Medical Journal. 2018;63:80-81.
10. Patra S, Agrawal N, Usha MK, Jayaranganath M. Common atrium with single ventricle: A rare combination of 2 uncommon complex congenital heart diseases. BMJ Case Rep. 2013:1-5. doi:10.1136/bcr-2013-200424.
11. Rastelli GC, Rahimtoola SH, Ongley PA, McGoon DC. Common atrium: Anatomy, haemodynamics, and surgery. J Thorac Cardiovasc Surg. 1968; 55:834-841.
12. 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;123:1214-7.