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UNIVENTRICULAR HEART IN TWO NIGERIAN ADOLESCENTS

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Abstract
Univentricular heart is an uncommon congenital heart disease. While early surgical intervention increases chances of survival, there are reports of individuals with uncorrected univentricular heart living into adulthood in western literature. In this report we present two cases of Nigerian adolescents, 12 and 16 years with different types of univentricular heart. Neither of them have had surgery, one of the cases died from transfusion related complication while the second one is alive and is being prepared for surgical intervention

Introduction

Univentricular heart is an uncommon congenital heart disease occurring in about 1-2 % of all congenital heart diseases.[1,2] It was found in 2.2% of all congential heart disease in a Nigerian study.[3] The congenital anomaly is defined as hearts in which the atrio-ventricular connections is completely or mostly to one ventricular chamber. The other ventricle is rudimentary and linked to the dominant ventricle via a bulboventricular foramen.[1] Univentricular heart is also known as single ventricle, triloculare, Holmes heart and biatriatum because of the various anatomical varieties.[1] The univentricular heart is primarily classified based on the morphology of the dominant ventricle (right or left ventricle), the atrioventricular connections and the ventriculo-aterial connections and the presence or absence of stenosis of the great vessels.

This complex malformation results from the abnormal development of the inlet and outlet portions of the embryonic heart tube which will form the definitive left and right ventricles respectively.[4] The prognosis in surgically uncorrected cases is reported to be poor with about 30% dying before their first birthday.[5] Early surgical correction is the preferred modality of treatment to prevent early mortality and ensure long term survival.[4]

There are reports of individuals with single ventricle living into adulthood in the Western literature.[5] There is however paucity of such reports in Nigeria. We report two cases of male adolescents who were diagnosed with univentricular hearts and had essentially uneventful life until demise as in the first case and is alive and well at 16 years of age, as in the second case.

KEY WORDS: Univentricular Heart, Polycythaemia, Exchange blood Transfussion

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Case report I

S.I, a 12 year old boy was first noticed to be cyanosed while crying at the age of two weeks. An echocardiogram done then confirmed single ventricle. The patient was neither seen subsequently in the clinic nor did he have any illness requiring admission. He attained normal developmental milestone during early childhood and started schooling at the age of 2 years. However his performance was well below average and he had to repeat classes even though he was reported to attend classes regularly. His clinical condition remained stable throughout childhood until the age of 11 years when he complained of weakness and worsening easy fatiguability. Polycythaemia was suspected because of suffused conjunctiva and the history of cyanosis from birth. Polycythaemia was confirmed by a haematocrit of 76%. He received partial exchange transfusion and got better. He received a second partial exchange four months later for a haematocrit of 67%. His symptoms subsided and remained clinically stable until he was referred to the paediatric cardiology unit for the first time at the age of 12 years for a possible surgical intervention.

In the cardiology clinic, he admitted to easy fatiquability. There was no history of breathlessness or squatting. He was not well grown for age, had a weight of 32 kg and height of 125 cm which represented 56 and70% of the expected respectively. He was centrally cyanosed and had suffused conjunctivae. There was grade 4 digital clubbing. The pulse rate was 82 beats per minute and regular, the blood pressure was 110/70 mmHg and he had left ventricular apical impulse. The apex beat was located in the 5th left intercostals space in the mid clavicular line. The first heart sound was normal while the second was single. There was a grade 3/6 pansystolic murmur loudest in the lower parasternal edge. The chest was clear of crepitations and the abdomen was full and soft without hepatomegaly. The chest radiograph showed cardiomegaly with a cardiothoracic ratio of 67%, normal pulmonary vascular markings and concave left upper heart border. (Fig I) The electrocardiogram revealed sinus rhythm, QRS axis was in the right upper quadrant, right and left atrial enlargement and right ventricular hypertrophy.(Fig II) The echocardiogram showed normal atrial and visceral situs and laevocardia. There was a common atrium which connected with the right sided coarsely trabeculated ventricle via a common atrioventricular valve which was mildly regurgitant. The interventricular septum extended to the crux and was thickened.(Fig III) The hypoplastic left ventricle was situated posteriorly, the great vessels arose from the main chamber. The mildly stenotic pulmonary artery was to the right and lying side by side the aorta. The pulmonary veins drained into the left side of the common atrium. The arch was left sided; there was no coarctation of the aorta or patent ductus arteriosus. There was good ventricular function.

The patient was being prepared for surgery abroad when he suddenly became symptomatic of polycythaemia for which he had a partial exchange. He apparently tolerated the procedure well initially, only to become suddenly very dyspnoeic 6 hours post exchange transfusion, lapsed into unconsciousness and eventually died.

Case report II

EU is a 16 year old boy who was first seen in paediatric cardiology unit at the age of 14 years with complaints of bluish discolouration of palms and soles and

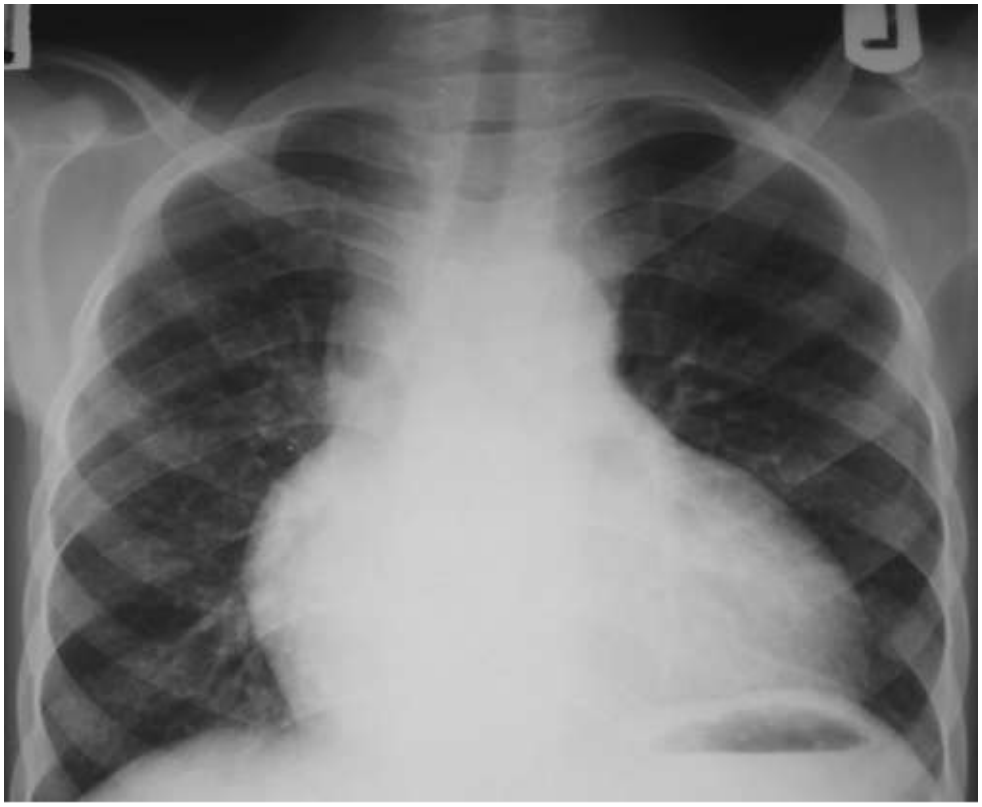


Fig I The chest radiograph showing cardiomegaly with concave pulmonary bay

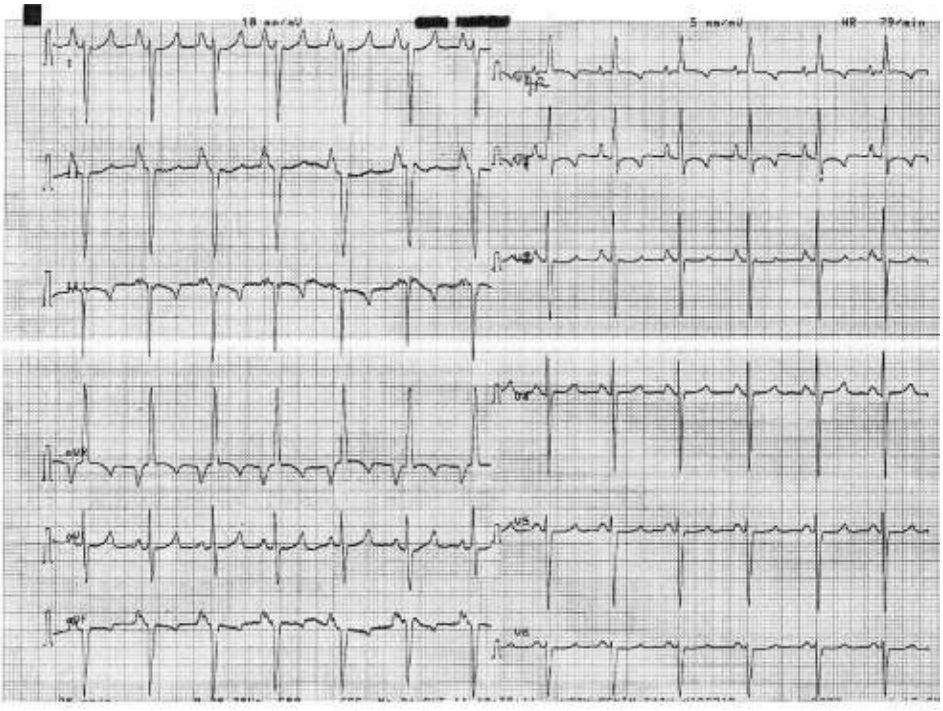
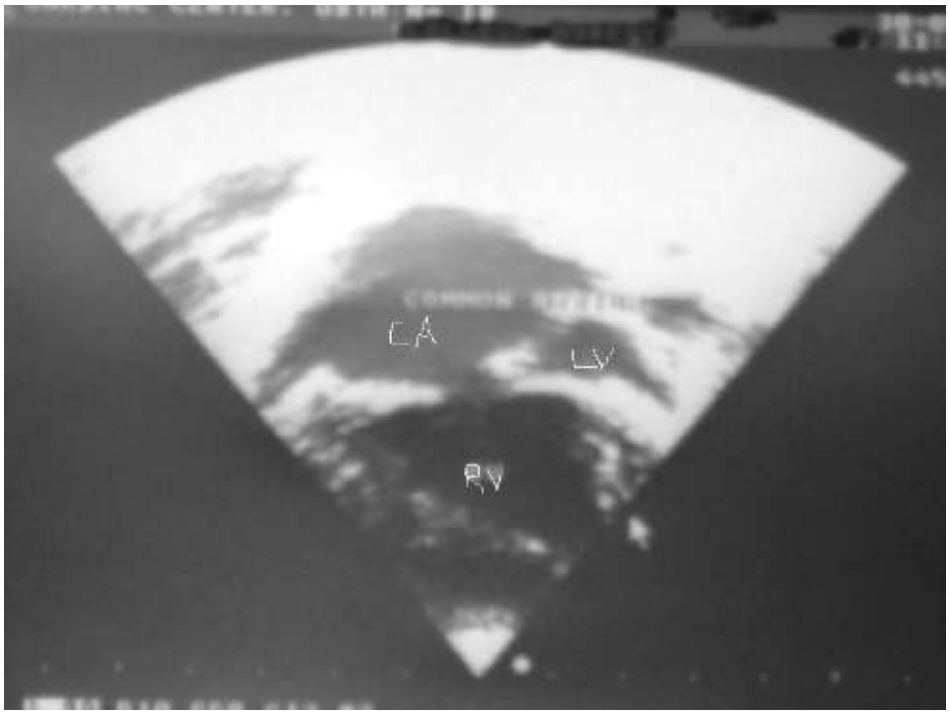


Fig II. The electrocardiogram showing right axis deviation, right ventricular and atrial hypertrophy



CA = common atrium, RV = right ventricle, LV = rudimentary left ventricle
Fig III. Subcostal view showing the common atrium (CA) with a common Atrioventricular valve emptying mainly into the right ventricle (RV) and the rudimentary ventricle (LV)

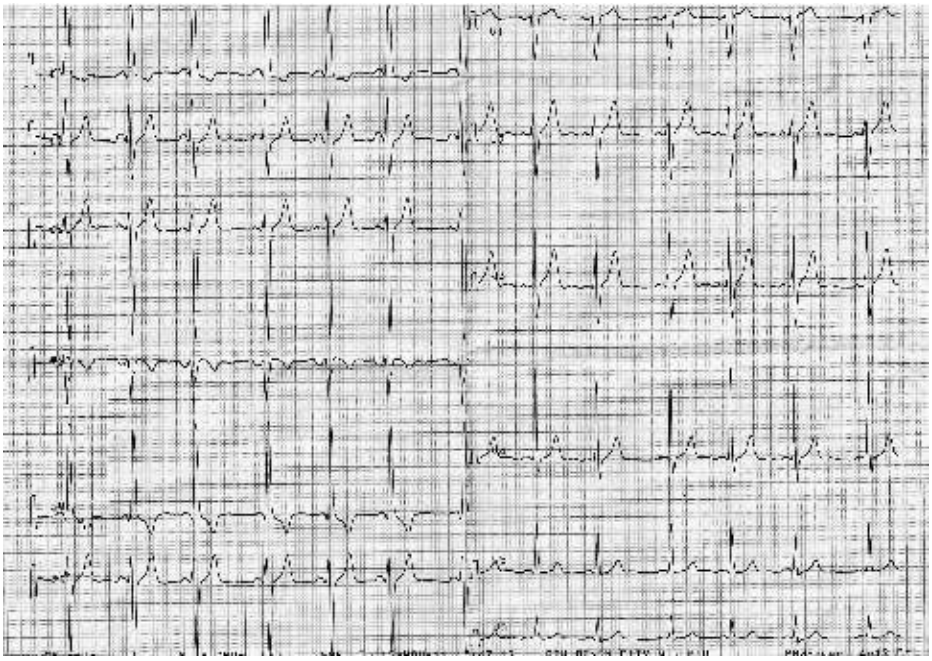
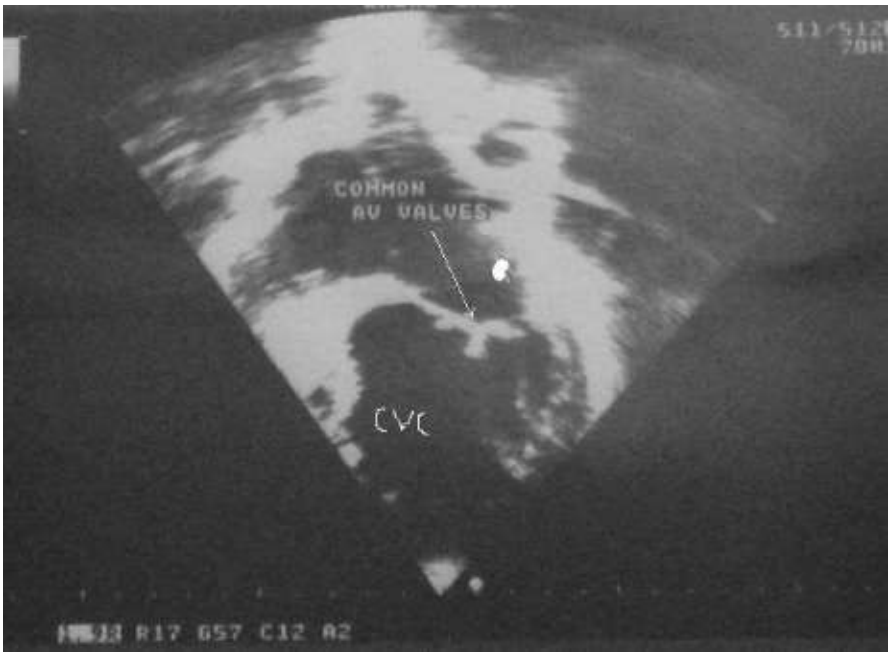


Fig IV. The ECG of a 16 year old boy showing left axis deviation and left ventricular forces in the precordial leads



CVC = common ventricular chamber

Fig V. Subcostal view showing common atrium, atrioventricular valve and common ventricular chamber.

recurring episodes of fast breathing and weakness, all of which have been occurring since birth. He achieved normal motor developmental milestones. He was an average student in class and in an appropriate class for his age. At presentation he was found to be asthenic with conjunctival suffusion, central cyanosis, grade 4 digital clubbing, the blood pressure was 100/70 mmHg and a grade two apical early systolic murmur and fixed splitting of the 2nd heart sound were heard. The chest radiograph showed a normal sized heart with a cardiothoracic ratio of 47% and normal pulmonary vascular markings. The ECG showed left axis deviation of – 47^{O}, predominant left ventricular (LV) forces in the precordial leads. (Fig IV). Echocardiography revealed a normal atrial situs, laevocardial, common atrium that is connected to a common ventricular chamber via a common mildly regurgitant atrio-ventricular valve (Fig V). The common ventricle has fine trabeculations. There is a smaller subaortic right-ventricular chamber located anteriorly, connected to both the common ventricular chamber and great vessels. The great vessels are transposed with the aorta anterior and to the left while the stenosed pulmonary artery is posterior and to the right. Both great vessels have bicuspid valves. The pulmonary venous drainage was unobstructive. The arch was left sided and there was no coarctation of the aorta. He was diagnosed with univentricular heart.

He has had two phlebotomies done at the age of 14 years and 17 years to correct polycythaemia of 72 and 74% respectively. Over the past 3 years he has not been on any routine medications or formal follow-up. Presently his oxygen saturation in room air is 74% (right thumb) and 42% (right second toe). His latest haematocrit is 56%. The patient is being prepared for surgery.

Discussion

The diagnosis of univentricular heart was made in both cases based on the echocardiogram finding of single-inlet left or right ventricles and the finding of rudimentary anterior ventricular chamber that connects to both the great arteries and the main ventricular chamber. The coarse trabeculations of the predominant ventricle as was in the first case and the fine pattern noted in the second case are in keeping with a right and left ventricular morphology respectively.[1] The extension of the interventricular septum to the crux is the usual finding in single ventricle of RV type as seen in the first case.[6] The transposed nature of the great vessels in both cases is the rule rather than the exception as shown in previous reports.[6,7] The RV type morphology of the first case is also shown in the ECG, with findings of extreme right axis deviation, tall R waves in the right chest leads and deep S waves in left chest leads as shown in Fig I. These are typical ECG changes noted in single ventricle of the RV type.[4] Similarly, the ECG findings of predominant LV forces in case II were supportive of the LV morphology.

Both patients did not have a dramatic presentation until later as adolescents when they presented with features of polycythaemia. This could have been due to the mild pulmonary stenosis that would have allowed just enough blood into the lungs and thus avoiding congestive cardiac failure from excess pulmonary blood flow or acidosis and cardiovascular collapse from marked decreased pulmonary blood flow. In previous reports of individuals with univentricular hearts living into adulthood, the consistent physiology was that of moderate pulmonary blood flow and transposed great vessels[7] as seen in the cases presented in this paper.[7]

There are previous reports of individuals with single ventricle living into adulthood and living a seemingly normal life.[6,8] Patients with single ventricle have been reported to carry pregnancy. Although a few have lost their lives or the babies during child birth,[9] most have been able to carry their pregnancies to term and had an uneventful delivery and post partum outcome.[10] It would appear that once the patients were in the second decade of life, they would be able to survive into fourth, fifth and sixth decades of life.

The patient in the first case report had an uneventful life until the age of eleven when he developed polycythaemia requiring partial exchange blood transfusion. This represents increasing level of desaturation either from pulmonary stenosis or worsening lung disease. There is no history to suggest progressive lung disease such as pulmonary arterial disease. We could not evaluate this since we did not have the benefit of serial echocardiograms or catheter studies. The pulmonary stenosis that was found on the last echocardiogram was not reported in the first echocardiogram done earlier in infancy. It is possible there was progressive pulmonary valvular stenosis that resulted in progressive desaturation later in life accounting for the sudden onset of polycythaemia. Although pulmonary stenosis is reported to be protective in many cases of single ventricle as it reduces

the pulmonary blood flow,[1,5] it may also be associated with mortality in its severe form where sudden cardiovascular collapse may occur. The sudden demise of the patient in the first case report after being stable post transfusion may have been complicated by transfusion related acute lung injury (TRALI). A condition caused by immune mediated pulmonary vascular permeability and acute pulmonary oedema.[11] Other possible causes include congestive heart failure from transfusion associated circulatory overload (TACO). There is a growing concern of the morbidity and mortality caused by TRALI and TACO in the transfusion medicine community.[11] In other cases reported previously, the death was unexplained.[5] It was however not possible to evaluate these complications in this child.

Conclusion

We have reported two cases (12 and 16year old boys) diagnosed with univentricular hearts and had uneventful lives until they were adolescents when they became symptomatic of polycythaemia requiring exchange transfusion. Although there are previous reports of individuals living into adulthood once they were in the second decade of life, the first case succumbed to complication of the exchange transfusion, while the other is alive.

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