Football actively, unhindered by any symptoms. However, 3 years later, he developed dyspnea on moderate exertion which gradually worsened over the next 3 years. There was associated cough productive of white sputum, with occasional hemoptysis. He subsequently developed bilateral leg swelling, orthopnea, paroxysmal nocturnal dyspnea, and occasional vomiting. He was not hypertensive, and there was no family history of hypertension or heart disease. He neither drank alcohol nor smoked cigarette.

He was first treated by a patent medicine dealer for about a year without improvement. Over the next 3–4 years, he sought medical attention in turn at a private hospital, a tertiary center, and an herbalist home before worsening symptoms prompted presentation at our center.

On examination, he was in respiratory distress, cachectic, centrally cyanosed, with grade 3 digital clubbing and bilateral pitting edema. His pulse rate was 120 beats/min, blood pressure 90/60 mmHg, jugular venous pulsation was raised, apex beat heaved at 6th left intercostal space anterior axillary line. There was a left parasternal heave, palpable P2, loud synchronous second heart sound, with a grade 3
presystolic murmur at the mitral area. His respiratory rate was 30 cycles/min; breath sounds were vesicular without adventitious breath sounds. He had hepatomegaly, but other systems were essentially normal.

Chest X-ray showed a biventricular cardiomegaly, mitralization of the left border of the cardiac silhouette and normal lung fields. The two-dimensional (2D)-echocardiography [Figure 1] showed grossly dilated left atrium and right ventricle, functional pulmonary regurgitation (Vmax 297 cm/s), pulmonary artery systolic pressure of 54 mmHg, a secundum ASD about 2 cm across with bidirectional shunting, flat interventricular septum in all the cardiac cycle; reversed transmitral diastolic flow velocities most likely due to pulmonary hypertension. Ejection fraction was 55%. A 12-lead electrocardiography [Figure 2] showed sinus rhythm, right axis deviation, right ventricular hypertrophy with strain, and inferior myocardial ischemia. Complete blood count revealed a hemoglobin level of 19.9 g/dl, packed cell volume of 61.9%, total white blood cell count of 6.7 x 10^9/L (neutrophils - 48%, lymphocytes - 52%) mean corpuscular volume 88.2 fl, mean corpuscular hemoglobin 28.3 pg, mean corpuscular hemoglobin concentration 32.1 g/dl, red cell distribution width 15.3 and a platelet count of 133 x 10^9/L.

He was treated with oral frusemide 40 mg daily, and sildenafil 40 mg daily with marginal improvement. Unfortunately, he was lost to follow-up.

**DISCUSSION**

ASD is the most common CHD in adults after bicuspid aortic valve. About 80% of ASDs are of the ostium secundum variety. Most cases of ASD become symptomatic in adolescence or adulthood with symptoms such as dyspnea on exertion, palpitations, cough and fatigue, as was the case with our patient. Late presentation is often generally due to the initial asymptomatic nature of ASD, but in low-income countries like Nigeria, other factors may be responsible. These factors include apathy for routine medical examinations, poverty, inadequate diagnostics and skilled workforce, and belief in “efficacy” of unorthodox medicine as portrayed by our patient.

ES is a rare complication of ASD, occurring in <5% of patients with ASD and its development probably requires genetic predisposition. While our patient was male, ES-complicated ASD predominantly affects females. ES is a multisystem disorder, characterized by progressive deterioration over time. Clinical presentation includes dyspnea, fatigue, hemoptysis, syncope, central cyanosis, digital clubbing, and heart failure in advanced stages. Our patient had virtually all these features. In general, patients with ASD are suspected to have ES when they have large, unrestricted defect and are cyanotic at rest. However not all ASD patients who develop cyanosis have ES. Cyanosis may be due to associated pulmonary stenosis or a prominent eustachian valve directing inferior vena cava flow to the left atrium through an ASD.

The diagnostic workup includes a painstaking history and physical examination, followed by investigations such as 2D or three-dimensional echocardiography, and cardiac magnetic resonance imaging if echo results are sub-optimal. Cardiac catheterization may be done to ascertain pulmonary vascular resistance and confirm right-to-left shunting. Electrocardiography, complete blood count, and chest X-ray constitute the ancillary investigations.

For treatment, three classes of drugs targeted at modifying endothelial dysfunction have been approved namely prostanoids, endothelin receptor antagonists, and phosphodiesterase type-5 inhibitors. Some studies demonstrated that these drugs lead to improvements in hemodynamics, exercise capacity, functional status, quality of life and survival in ES patients. More recently carefully selected patients with ASD complicated by ES were reported to have benefitted from corrective repair after prolonged advanced therapy with oral sildenafil, oral bosentan, or intravenous epoprostenol.

Nevertheless, most Eisenmenger patients die from sudden cardiac death, congestive heart failure, hemoptysis, cerebral abscesses, thromboembolic events, and complications during pregnancy or due to noncardiac surgery.

**CONCLUSION**

ES seen in our patient is a rare complication of ASD which could have been prevented if he had presented to our center earlier. Late presentation can lead to the development of
ES, which precludes surgical repair, except in very few carefully selected cases.

Acknowledgment
The authors would like to thank Dr. Ejim EC for producing the 2D echocardiography.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

REFERENCES
1. Eisenmenger V. The Congenital defect of the chamber walls of the heart. Z Klin Med 1897;32:1-28.
2. Steele PM, Fuster V, Cohen M, Ritter DG, McGoon DC. Isolated atrial septal defect with pulmonary vascular obstructive disease – Long-term follow-up and prediction of outcome after surgical correction. Circulation 1987;76:1037-42.
3. Baumgartner H, Bonhoeffer P, De Groot NM, de Haan F, Deanfield JE, Galie N, et al. ESC Guidelines for the management of grown-up congenital heart disease (new version 2010). Eur Heart J 2010;31:2915-57.
4. Ejim EC, Anisiuba BC, Ike SO, Essien IO. Atrial septal defects presenting initially in adulthood: Patterns of clinical presentation in Enugu, South-East Nigeria. J Trop Med 2011;2011:251913.
5. Daliento L, Somerville J, Presbitero P, Menti L, Brach-Prever S, Rizzoli G, et al. Eisenmenger syndrome. Factors relating to deterioration and death. Eur Heart J 1998;19:1845-55.
6. Webb G, Gatzoulis MA. Atrial septal defects in the adult: Recent progress and overview. Circulation 2006;114:1645-53.
7. Dimopoulos K, Inuzuka R, Goletto S, Giannakoulas G, Swan L, Wort SJ, et al. Improved survival among patients with Eisenmenger syndrome receiving advanced therapy for pulmonary arterial hypertension. Circulation 2010;121:20-5.
8. Galìè N, Beghetti M, Gatzoulis MA, Granton J, Berger RM, Lauer A, et al. Bosentan therapy in patients with Eisenmenger syndrome: A multicenter, double-blind, randomized, placebo-controlled study. Circulation 2006;114:48-54.
9. Kim YH, Yu JJ, Yun TJ, Lee Y, Kim YB, Choi HS, et al. Repair of atrial septal defect with Eisenmenger syndrome after long-term sildenafil therapy. Ann Thorac Surg 2010;89:1629-30.