INTRODUCTION

Ebstein's anomaly is a complex congenital heart disease due to failure of delamination of the tricuspid valve leaflets from the interventricular septum, resulting in adherence of the leaflets to the underlying myocardium. This results in a wide variety of cardiac abnormalities, including apical and posterior displacement of the dilated tricuspid valve annulus, dilation of the atrialized portion of the right ventricle, fenestrations, redundancy, and tethering of the anterior leaflet of the tricuspid valve. The malformed tricuspid valve is usually regurgitant and rarely be stenotic. The clinical manifestations of Ebstein's anomaly in the adult depend on several factors, including the extent of tricuspid valve leaflet distortion, degree of tricuspid regurgitation, right atrial pressure, and presence of a right-to-left atrial level shunt. Adult patients with congenital heart disease may have abnormal glucose metabolism or elevated circulating levels of inflammatory cytokines. We are reporting the first case where diabetes ketosis being the presentation, before the diagnosis of Ebstein's anomaly.

CASE REPORT

A 35-year male patient was admitted with generalized fatigue and chest pain for the last ten days. He gave the history of palpitation, effort intolerance and dyspnoea of New York Heart Association (NYHA) class II severity for the last 5-6 years though never showed to any physician. These symptoms were increased in the last 10 days. He was a non-smoker and non-alcoholic and there was no family history of diabetes or heart disease. On general physical examination, he had engorged jugular venous pressure with prominent 'A' and 'V' wave without any presence of cyanosis, edema or clubbing. Cardiac system examination revealed the presence of wide split first heart sound with prominent tricuspid component, single second heart sound and soft grade II/VI early decrescendo systolic murmur at lower parasternal area. Per abdominal examination revealed mild hepatomegaly without any hepatic pulsation. Respiratory and other system examinations were within normal limit. Blood investigations revealed normal complete blood count (Hemoglobin 13.2 gm/dl; total leukocyte count 8600/cmm; platelets-235000/cmm), normal renal function test (blood urea -34 mg/dl; serum creatinine-1.1 mg/dl) and liver function tests. His random blood sugar at presentation was 512 mg/dl and urine showed positive test for ketone bodies. Arterial blood gas was sent to rule out diabetic ketoacidosis and it was found to be compensatory metabolic acidosis (pH-7.36; HCO3-16 mmol/L; pCO2-35 mm of Hg). He had high fasting (136 mg/dl) and post-prandial (260 mg/dl) blood sugar. Chest X-ray revealed right atrial enlargement and cardiomegaly (cardio-thoracic ratio of 55%). Electrocardiogram revealed tall P wave, prolonged PR (200 ms) and left axis deviation without any delta wave. Transthoracic echocardiography revealed the TTE apical displacement of septal leaflet of tricuspid valve by 37.6 mm (20 mm/m2 of body surface area), giant right atrium, and displacement of ventricular septum towards left ventricular cavity (Figure 1) without presence of any right to left shunt. Doppler echocardiography revealed low pressure organic TR begins at the level of displaced septal and posterior leaflets of tricuspid valve and courses through the atrialized right ventricle into the atrium.
right atrium proper (Figure 2). He was managed conservatively with regular insulin infusion followed by split doses of long acting insulin and diuretic therapy for right ventricular dysfunction.

**DISCUSSION**

Ebstein's anomaly of the tricuspid valve occurs as an isolated defect, with other forms of congenital heart disease such as transposition of the great arteries or tetralogy of Fallot or, rarely, in association with extra cardiac malformations. Major extra cardiac changes most often involved the craniofacial region, central nervous system, and limbs. Whereas, type I diabetes mellitus is rarely associated with congenital heart defects like Ellis-van Creveld syndrome, ventricular septal defect and patent ductus arteriosus. Several studies had showed the surprising prevalence of abnormal glucose metabolism in congenital heart disease, which proved to be about 10 times higher than their general peers. Because all neurohormonal activities were increased in congenital heart disease groups, the heart failure related mechanism contributes to the high prevalence of insulin resistance. Congenital heart disease like Ebstein's anomaly often have right-sided heart failure that causes liver congestion; therefore, secondary liver dysfunction due to fatty liver and/or hemodynamic abnormalities caused by congestive heart failure may be associated with abnormal glucose metabolism or diabetes mellitus.

In this case, the patient had right ventricular dysfunction and features of congestive heart failure. So, the decompensated heart failure might be the reason for precipitation of diabetes ketosis in previously undiagnosed case of both diabetes mellitus and Ebstein's anomaly. Though recently, GATA 6 mutation is associated with childhood diabetes, pancreatic agenesis and congenital heart defects, but nowhere mentioned that diabeted ketoacidosis was associated with Ebstein's anomaly.

This report demonstrated that complex congenital heart disease like Ebstein's anomaly can be associated with diabetes mellitus and diabetes ketoacidosis could be the presentation. So, abnormal glucose metabolism should be considered, diagnosed and treated when we manage complex congenital heart disease like Ebstein's anomaly.

**REFERENCES**


