Journal of Young Pharmacists, 2024; 16(3):594-596.
https://www.jyoungpharm.org

Case Report

Ebstien Anomaly with Cardiomegaly in a Paediatric Patient: Case Report

Tahameen K kurubanavar, Bibi Khuteja Jangliwale, Arnab Sarkar, Agadi Hiremath Viswanatha Swamy, Prajnashree Acharya, Sanatkumar Bharamu Nyamagoud*

Department of Pharmacy Practice, KLE College of Pharmacy, Vidyanagar, Hubballi, A Constituent Unit of KLE Academy of Higher Education and Research, Belagavi, Karnataka, INDIA.

ABSTRACT

Ebstien anomaly is a rare heart problem present at birth and has a varied clinical course, it's a congenital heart defect of the tricuspid valve. We hereby describe a 13-year-old child, was presented with history of Ebstien anomaly with Ostium secundum Atrial Septal Defect (OS ASD) was diagnosed at 1 year of age, but surgical correction was not done in early decade due to financial problem. At admission, child was in shock and saturation was not maintaining, Echocardiogram examination revealed Congenital Heart Disease (CHD), Ebstien anomaly, RA/RV dilated, atrialised Right Ventricle (RV), Septal Tricuspid Leaflet (STL) displaced to the origin distance 5.6cm was noted, Grade 2+ Tricuspid Regurgitation (TR), Pulmonary Artery Systolic Pressure (PASP)-28 mmHg, no Pulmonary Arterial Hypertension (PAH), normal Biventricular function. Child needed immediate surgical correction. Child was discharged with referral letter to higher centre.

Keywords: Ebstien anomaly, Atrial septal defect, Tricuspid regurgitation, Cone repair, Coarctation of aorta, congenital heart disease.

Correspondence:
Dr. Sanatkumar Bharamu Nyamagoud
Assistant Professor, Department of Pharmacy Practice, KLE College of Pharmacy, Vidyanagar, Hubballi, A Constituent Unit of KLE Academy of Higher Education and Research, Belagavi, Karnataka, INDIA.
Email: dr.sanathnyamagoud@gmail.com

Received: 20-02-2024;
Revised: 03-04-2024;
Accepted: 05-05-2024.

INTRODUCTION

Ebstien anomaly is a relatively uncommon congenital heart disease. It is defined as apical tricuspid valve displacement, leading Tricuspid Regurgitation (TR), right ventricle function reduction and right ventricle and right atrium dilation. Teratology of Fallot (TOF) is most commonly associated with Coarctation of Aorta (CoA).[1]

CASE REPORT

A 13-year-old Indian boy presented to an outpatient clinic with fever, vomiting, neck pain and headache since past 4 days. His mother reported that child was apparently well 4 days back, child is a known case of Ebstien anomaly with Ostium Secundum Atrial Septal Defect (OS ASD) with TR, now presented with fever since 4 days insidious in onset intermittent to low grade not associated with rigor and rashes, aggravating factors not relived on taking medication, history of vomiting since 4 days non-foul-smelling, history of neck pain since 4 days non-radiating, history of intermittent headache since 4 days, No history of sore throat, dry skin lesion, chest pain, palpitation, forehead sweating, any lesion on finger tips, abdominal pain, distention, loose stools, altered sensorium and seizures. He was 3rd born to non-consanguineous mother and father, he was born in home FTVD birth weight is 2.5 kg, mother was not receiving any medication during her antenatal period, ANC scan was normal. Child developed appropriate for the age. There was no family history of connective tissue disorder in the family. Child belongs to the lower socioeconomic status. At presentation, he was febrile (104.0°F), tachypnoeic with blood pressure lies between 95th. Saturation was 87% on room air in the index finger. Weight of the child was <3rd percentile per (World Health Organisation) standards; height was between 3rd-10th percentile and BMI was <3rd percentile.

Head to toe examination revealed normal fundus, on chest apical impulse seen, abdomen 1x1 cm café-au-lait spots, cyanosis and grade 2 clubbing in upper limbs and cyanosis and clubbing of nails (Figure 1). CVD examination reveals normal shape, no chest deformity and bulge, tracheal position is normal, apical impulse felt at 7th ICS lateral to MCL, neck veins are not engaged. On palpation apical impulse felt at 6th ICS 2nd lateral to MCL shifted downwards and outcomes hyperdynamic apical impulse more than one ICS. On auscultation S1S2 positive, systolic murmurs grade IV and split of S2. Respiratory examination reveals central trachea, B/L chest raise equal, bilateral air entry equal and normal vesicular breath sounds heard. Complete blood count was within normal range, serum test reveals sodium and potassium were reduced, liver function test was normal, His chest radiograph

DOI: 10.5530/jyp.2024.16.74

Copyright Information :
Copyright Author (s) 2024 Distributed under Creative Commons CC-BY 4.0

Publishing Partner : EManuscript Tech. [www.emanuscript.in]

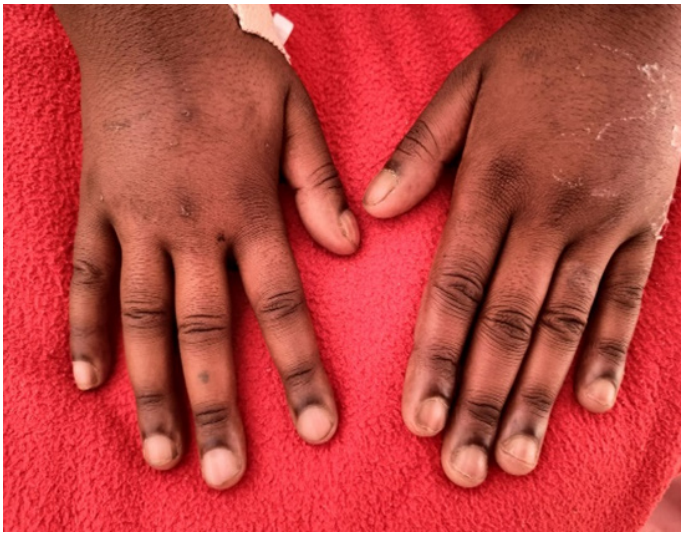

Figure 1: Grade 2 clubbing in upper limbs.

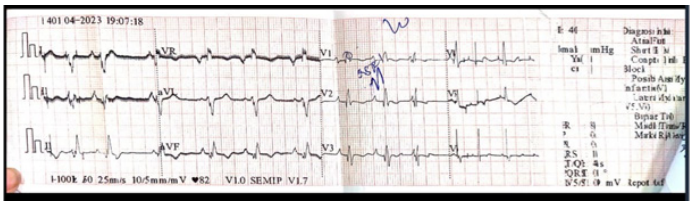

Figure 2: Electrocardiogram reveals bifascicular block (RBBB) P pulmonale, T inversion in lead 3, avF, v1-v6 widen QRS in V1 Rsr' pattern.

revealed cardiomegaly. In electrocardiogram Reveals Bifascicular Block (RBBB) P pulmonale, T inversion in lead 3, avF, v1-v6 widen QRS in V1 Rsr' pattern seen (Figure 2). Echocardiogram (Echo) revealed CHD, Ebstien anomaly, RA/RV dilated, arterialised RV, STL displaced to the origin distance 5.6 cm was noted, Grade 2+ TR PASP-28 mmHg, no PAH, normal B/V function (Figure 3). A 20 mL/kg IVF NS bolus given; 0.1 mg/kg/min adrenaline infusion started. i/v/o CHD 6 µg/kg/min dobutamine started. Oxygen saturation was maintained by oxygen mask. After 1 hr BP improved 130/110 mmHg hence adrenaline was reduced to 0.07 mcg/kg/min and hence dobutamine increased to 10 µg/kg/min, Injection PCT 25 mL was given immediately and child was given Injection Ceftriaxone and gradually inotropes were tapered and stopped and monitored for vitals, BP maintaining between 95th+12 mmHg centile, started on antihypertensive nifedipine 0.3 mg QID. Patient referred to the higher authorities because child needed the surgical correction.

DISCUSSION

Ebstien anomaly is a rare heart problem present at birth, it's a congenital heart defect of the tricuspid valve, it occurs 1 in 2,00,000 lives births and accounts for less than one percent of all CHD.[1] By anatomically, EA is defined as the displacement of the TV's posterior and septal leaflets by more than 8 mm/m^2 of body surface area in relation to the location of the anterior leaflets of the mitral valve.[2] Nevertheless, the cause of EA is

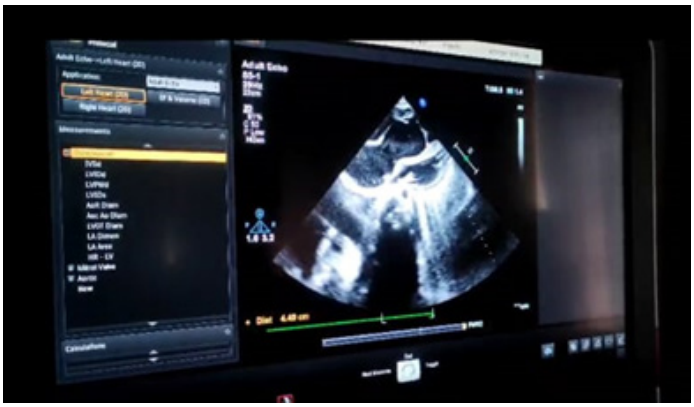

Figure 3: STL displaced to the origin distance 5.6 cm was noted.

uncertain. Mutation in the transcription factor NKX2.5, 10p 13-p14 deletion, 1p34.3-p36.11 deletion, these are genetic factors which are included in Ebstien anomaly, morphological defects include TV leaflets failure which resulting in attachment of the leaflets to the underlying myocardium. This causes a variety of abnormalities, including apical and posterior displacement of the dilated tricuspid valve annulus and dilation of the right ventricular Arterialised portion. Ebstien's anomaly can range from symptomatic newborns to non-symptomatic adults. Cases of Ebstein's anomaly have been reported in newborns, young children, teens and adults.[3]

The clinical presentation after the neonate's time of life has a better prognosis than a case diagnosed in early life.[4] Adults may have fewer anatomical anomalies due to high compliance of the volume overload in the right chamber. Patients are generally symptomatic for a longer period of time. The average age of diagnosis in children is 13 years and in adults it is 20-30 years.[4,5] At the time of diagnosis, the vast majority of adult patients have these New York Heart Association (NYHA) class I and class II characteristics, Arrhythmias are most prevalent in children and adults.[6]

Right-to-A left shunt can cause chronic hypoxia and exertional cyanosis and Children are more likely to suffer from cyanosis. Cerebral vascular embolic events are attributed to paradoxical embolization or the occurrence of atrial fibrillation in approximately 8%-10% of cases.[7]

First line diagnostic tool is Transthoracic echocardiography. Transoesophageal echocardiography and cardiovascular magnetic resonance are second-line procedures. The former is particularly useful in Tricuspid Valve (TV) assessment, while the latter is useful in right-sided chamber volume calculation and myocardial characterization. Cardiovascular magnetic resonance detects posterior leaflet and extra-cardiac abnormalities better than Transthoracic echocardiography, but Transthoracic echocardiography more often detects small septal transmission. Echocardiography and Cardiovascular magnetic resonance are both recommended for evaluation and unoperated patients.[3,4]

In patients with Ebstien anomaly, it is recommended to have a clinical follow-up examination at least once a year, though the monitoring strategy can be adjusted to the patient's specific features. Main clinical symptoms known to affect prognosis in Ebstien anomaly are HF, arrhythmias and cyanosis, which can be addressed through a multidisciplinary approach of monitoring, prevention and treatment.[8]

Medical therapy's impact on EA patients has yet to be thoroughly examined but it can be managed by observation, medical or surgical management. Digoxin and beta-blockers are used to reduce induced ventricular dysfunction and tachycardia. Antiarrhythmic class IC and III drugs may assist in the abnormal heart rhythm management.[9] however, sotalol and amiodarone because of their known proarrhythmic risk and side effects, they should be avoided as long-term therapy. In the matter of AF and right-to-left shunt oral anticoagulation therapy may be applicable. When there is a BBB and either left ventricle or right ventricle condition cases, cardiac resynchronisation therapy can be used.[10]

In EA, TV is usually inept and occasionally stenotic. Fenestrations in the anterior leaflet may contribute to tricuspid regurgitation. Anterior leaflet may have fenestrations that contribute to tricuspid regurgitation and right ventricular outflow tract obstruction caused by fibro-muscular attachment. In surgical patients, the clinical outcome CR outperformed previous repair techniques in terms of residual tricuspid regurgitation, freedom of re-operation and durability.[11] The benefits of CR include the ability to grow and maintain native tissue, as well as the need for long-term anticoagulation therapy is eliminated.[4]

Patients with an ASD or PFO, as well TR mild-moderate or moderate-mild may be benefited from transcatheter closure to prevent the development of paradoxical emboli and improve cyanosis.

CONCLUSION

Ebstien anomaly is extremely rare and fascinating heart defect at birth disease (CHD) with variable course. As a result, precise understanding of the various presentations, prompt diagnosis and appropriate management options are required. Immediate correction of cyanosis or else it leads to respiratory failure or death. As a result, surgical correction is crucial in this condition.

ACKNOWLEDGEMENT

The Authors are thankful to the VC, Registrar and Dean of Pharmacy, KAHER, Belagavi. We would also like to thank patient, medical and hospital staff of VGH, Hubballi.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

ABBREVIATIONS

ASD: Atrial Septal Defect; **CHD:** Congenital Heart Disease; **TR:** Tricuspid Regurgitation; **STL:** Septal Tricuspid Leaflet; **RV:** Right Ventricle; **PASP:** Pulmonary Artery Systolic Function; **OS:** Ostium, Secundum; **RBBB:** Right Bundle Branch Block.

REFERENCES

1. Acharya P, Ang JR, Gitler B. Ebstein anomaly with QRS fragmentation on electrocardiogram. J Investig Med High Impact Case Rep. 2017;5(1):232470961668871. Available from: http://dx.doi.org/10.1177/2324709616688710
2. Manuel V, Morais H, Magalhães MP, Nunes MAS, Leon G, Ferreira M, et al. Ebstein's anomaly in children: A single-center study in Angola. Rev Port Cardiol. 2015;34(10):607-12. Available from: http://dx.doi.org/10.1016/j.repc.2015.03.015
3. Krieger EV, Valente AM. Diagnosis and management of ebstein anomaly of the tricuspid valve. Curr Treat Options Cardiovasc Med. 2012;14(6):594-607. Available from: http://dx.doi.org/10.1007/s11936-012-0209-2
4. Pasqualin G, Boccellino A, Chessa M, Ciconte G, Marcolin C, Micaglio E, et al. Ebstein's anomaly in children and adults: multidisciplinary insights into imaging and therapy. Heart. 2023;heartjnl-2023-322420. Available from: http://dx.doi.org/10.1136/heartjnl-2023-322420
5. Herrera-Bello H, Ávila-Vanzzini N, Fritche-Salazar JF, Kuri-Alfaro J, Gaxiola-Macias MBA, Cossio-Aranda JE. Clinical spectrum and long-term outcome of 183 cases of ebstein's anomaly, experience of Mexican population. Arch Med Res. 2020;51(4):336-42. Available from: http://dx.doi.org/10.1016/j.arcmed.2020.03.008
6. Attie F, Rosas M, Rijlaarsdam M, Buendia A, Zabal C, Kuri J, et al. The adult patient with Ebstein anomaly. Outcome in 72 unoperated patients. Medicine (Baltimore). 2000;79(1):27-36. Available from: http://dx.doi.org/10.1097/00005792-200001000-00003
7. Godart F. Atrial right-to-left shunting causing severe hypoxaemia despite normal right-sided pressures. Report of 11 consecutive cases corrected by percutaneous closure. Eur Heart J. 2000;21(6):483-9. Available from: http://dx.doi.org/10.1053/euhj.1999.1944
8. Singh DP, Hussain K, Mahajan K. Ebstein Anomaly and Malformation. StatPearls Publishing; 2023.
9. Kron IL, Roeser ME. Management of Ebstein's anomaly. Ann Cardiothorac Surg. 2017;6(3):266-9. Available from: http://dx.doi.org/10.21037/acs.2017.05.03
10. Kong NW, Upadhyay GA. Cardiac resynchronization considerations in left bundle branch block. Front Physiol. 2022;13. Available from: http://dx.doi.org/10.3389/fphys.2022.962042
11. Burri M, Mrad Agua K, Cleuziou J, Beran E, Nagdyman N, Kühn A, et al. Cone versus conventional repair for Ebstein's anomaly. J Thorac Cardiovasc Surg. 2020;160(6):1545-53. Available from: http://dx.doi.org/10.1016/j.jtcvs.2020.05.032

Cite this article: Jangliwale BK, kurubanavar TK, Sarkar A, Nyamagoud SB, Dayana BM. Ebstien Anomaly with Cardiomegaly in a Paediatric Patient: Case Report. J Young Pharm. 2024;16(3):594-6.