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Ebstein's Anomaly in a Young Nigerian Lady

L'anomalie d'Ebstein chez une jeune Nigériane

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ABSTRACT

BACKGROUND: Ebstein's anomaly (EA) is a rare congenital heart disease with a wide spectrum of presentation from the womb to adulthood characterized by apical displacement of tricuspid valve leaflets into the right ventricle. This is due to defects in embryological processes of uncertain etiology.

CASE SUMMARY: A 28-year-old woman who presented with palpitations from childhood occurring in short paroxysms daily and limiting her from strenuous activities. Physical examination findings revealed a small stature with no associated birth defects. The cardiovascular system revealed tachycardia, a small volume irregular pulse with a split S2 heart sound. ECG showed premature atrial complexes, fragmented QRS with Sodi-Pallares sign, and echocardiographic features in keeping with atrialization of right ventricle with apical displacement of septal leaflet of the tricuspid valve into the right ventricle in keeping with EA. The patient was placed on anti-arrhythmics and anticoagulants.

CONCLUSION: The diagnosis of EA in adults is often delayed in low-resource settings for multifaceted reasons. However, medical therapy offers a conservative measure to preserve life while surgical correction is advocated for the majority of cases when picked up early. **WAJM 2024; 41 (6): 727 - 734**

KEYWORDS: Ebstein's Anomaly, Case Report, Literature Review

RÉSUMÉ

CONTEXTE: L'anomalie d'Ebstein (AE) est une cardiopathie congénitale rare caractérisée par le déplacement apical des feuillets de la valve tricuspide dans le ventricule droit. Cette affection se manifeste par un large éventail de symptômes, de la vie fœtale à l'âge adulte, et serait due à des défauts dans les processus embryologiques, bien que son étiologie exacte reste incertaine.

RÉSUMÉ DU CAS: Une femme de 28 ans s'est présentée avec des palpitations depuis l'enfance, se produisant en courtes paroxysmes quotidiens et la limitant dans les activités physiques intenses. L'examen physique a révélé une petite stature sans malformations associées. L'évaluation cardiovasculaire a indiqué une tachycardie, un pouls irrégulier de faible volume et un dédoublement du deuxième bruit du cœur (B2). Un ECG a montré des complexes atriaux prématurés, des complexes QRS fragmentés avec un signe de Sodi-Pallares, et des résultats échocardiographiques cohérents avec une atrialisation du ventricule droit et un déplacement apical du feuillet septal de la valve tricuspide, confirmant un diagnostic d'AE. La patiente a été prise en charge avec des antiarythmiques et des anticoagulants.

CONCLUSION: Dans les milieux à faibles ressources, le diagnostic de l'AE chez l'adulte est souvent retardé en raison de divers défis. Bien que la prise en charge médicale offre une approche conservatrice pour prolonger la vie, une intervention chirurgicale précoce est recommandée dans la plupart des cas pour obtenir de meilleurs résultats. **WAJM 2024; 41 (6): 727 - 734**

MOTS-CLÉS: Anomalie d'Ebstein, Rapport de cas, Revue de la littérature

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