



ORIGINAL ARTICLE

Ebstein's anomaly in children: A single-center study in Angola



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KEYWORDS

Congenital heart disease;
Congenital heart surgery;
Valve lesion;
Tricuspid valve;
Cone reconstruction

Abstract

Introduction and Objective: Ebstein's anomaly is a rare complex congenital heart defect of the tricuspid valve. We aimed to describe the frequency, clinical profile, and early and short-term post-operative results in patients under the age of 18 years operated for this anomaly in a tertiary center in Angola.

Methods: A retrospective cross-sectional study was conducted over a period of 37 months. We analyzed all patients diagnosed with congenital heart defects.

Results: Of the 1362 patients studied, eight (0.6%) had Ebstein's anomaly; six patients (75%) were female. Mean age was 69 ± 59 months. Five patients were in NYHA functional class III or IV. Mean cardiothoracic index was 0.72. Seven patients (87.5%) had severe tricuspid regurgitation and five (62.5%) had another associated congenital heart defect. All patients were operated: two had complications and one (12.5%) died in the early post-operative period. The mean follow-up time was 1.22 ± 0.6 years, and mortality during follow-up was 12.5% (n=1). At the end of the study, of the five patients in whom cone reconstruction was performed, four (80%) were in functional class I. Mean cardiothoracic index decreased to 0.64. Three patients had mild and two had moderate tricuspid regurgitation. The patient who underwent cone reconstruction and a Glenn procedure was in functional class I.

Conclusion: The frequency of Ebstein's anomaly was similar to that in other centers. Cone reconstruction was viable in the majority of patients, with good early and short-term results.

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PALAVRAS-CHAVE

Cardiopatía congénita;
 Cirurgia cardíaca congénita;
 Lesão valvular;
 Válvula tricúspide;
 Reconstrução em cone

Anomalia de Ebstein em crianças: estudo unicêntrico em Angola**Resumo**

Introdução e Objectivos: A anomalia de Ebstein é uma cardiopatía congénita complexa e rara da válvula tricúspide. Descrever a frequência, o perfil clínico e os resultados pós-operatórios imediatos desta anomalia em crianças e adolescentes.

Métodos: Estudo transversal retrospectivo no único centro com tratamento integrado em cardiopatías congénitas em recém-nascidos, crianças e adolescentes (<18 anos de idade) em Angola, num período de 37 meses. Foram analisados todos os doentes com diagnóstico de cardiopatía congénita. O diagnóstico foi feito com base no exame clínico e dados da ecocardiografia transtorácica e Doppler. Os ecocardiogramas foram realizados pelos Cardiologistas Pediátricos numa máquina Vivid 7 G.E.

Resultados: Foram analisados 1362 ecocardiogramas sequenciais. Oito doentes (0,6%) tinham anomalia de Ebstein, destes 6 (75%) eram do género feminino. As idades variaram entre 4 a 168 meses, a média foi de 69 ± 59 meses. Cinco estavam em classe funcional III ou IV da NYHA. A média do índice cardiotorácico foi de 0,72. Sete doentes (87,5%) tinham regurgitação tricúspide grave e 5 (62,5%) tinham outra cardiopatía congénita associada. Todos os doentes foram submetidos a intervenção cirúrgica. Dois tiveram complicações no pós-operatório imediato. A mortalidade pós-operatória foi de 12,5% (n=1).

Conclusão: A frequência da Anomalia de Ebstein encontrada foi semelhante a de outros Centros. A técnica de Cone foi viável na maior parte dos pacientes. A mortalidade pós-operatória imediata foi baixa.

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List of abbreviations

| | |
|------|-----------------------------|
| ASD | atrial septal defect |
| CHD | congenital heart defect |
| CR | cone reconstruction |
| CTI | cardiothoracic index |
| EA | Ebstein's anomaly |
| MBT | modified Blalock-Taussig |
| NYHA | New York Heart Association |
| PAH | pulmonary artery hypoplasia |
| PDA | patent ductus arteriosus |
| RV | right ventricular |
| TV | tricuspid valve |
| WPW | Wolff-Parkinson-White |

Introduction

Ebstein's anomaly (EA) is a rare complex congenital heart defect (CHD) of the tricuspid valve (TV) first described by Wilhelm Ebstein in 1866.¹ It occurs in about 1 per 200000 live births and accounts for less than 1% of all CHDs.²⁻⁸ The anatomical basis of EA consists in displacement by more than 8 mm/m² body surface area of the septal and posterior leaflets of the TV in relation to the position of the mitral valve anterior leaflet.^{4,6} The genetic etiology is unknown.⁵ The usual clinical presentation is cyanosis, heart failure, arrhythmia and sudden death.^{3,8} The severity and onset of symptoms depend on the degree of

displacement of the TV leaflets.³ Transthoracic echocardiography is the gold standard for diagnosis of this entity.^{4,6} EA may coexist with other CHDs, genetic syndromes, or Wolff-Parkinson-White (WPW) syndrome.^{2-4,9} Surgical management of EA depends on the patient's age, the clinical presentation and association with other CHDs, and therefore the surgical approach should be individualized.^{9,10} Ebstein's anomaly is classified among the category of CHDs that requires early and specific health care.^{7,8} In this context, the present study aimed to describe the experience of a tertiary center in a developing country in managing this complex disease in patients less than 18 years old.

Methods

Based on the records of the Cardiothoracic Center of Clínica Girassol, previously described,¹¹ a retrospective study was performed that included all patients with CHDs over a period of 37 months (March 29, 2011 to April 29, 2014). Data on clinical presentation, chest X-ray, electrocardiography, echocardiographic studies and surgical reports were analyzed. Transthoracic echocardiography and Doppler studies was performed by pediatric cardiologists using a GE Vivid 7 system (GE Healthcare, Milwaukee, WI, USA) with multiple frequency probes (3S-RS, 4C-RS and RS-6S) in accordance with the recommendations of the American College of Cardiology/American Heart Association.¹² Only the first echocardiogram was validated. A diagnosis of EA was made when a displacement of the septal and posterior leaflets of the TV was observed of more than 8 mm/m² body

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