Rev Port Cardiol. 2015;34(10):607–612

Revista Portuguesa de Cardiologia
Portuguese Journal of Cardiology
www.revportcardiol.org

ORIGINAL ARTICLE

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

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Received 6 February 2015; accepted 21 March 2015
Available online 29 September 2015

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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http://dx.doi.org/10.1016/j.repc.2015.03.015

PALAVRAS-CHAVE
Cardiopatia 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 cardiopatia 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 retrospetivo no único centro com tratamento integrado em cardiopatias 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 cardiopatia 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 cardiopatia 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.

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 200 000 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/m^2 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/m^2 body