

# TRANS-CATHETER CLOSURE OF LARGE SECUNDUM ATRIAL SEPTAL DEFECTS IN PATIENTS WITH EBSTEIN'S MALFORMATION OF THE TRICUSPID VALVE: A REPORT OF TWO CASES

<sup>1</sup>Mohammed T Numan, <sup>2</sup>Sulafa KM Ali

*Pediatric Cardiology Section, <sup>1</sup>Department of Cardiology & Cardiovascular Surgery, Hamad Medical Corporation, Doha, Qatar;*

*<sup>2</sup>Pediatric Cardiology Department, Sudan Heart Centre, Khartoum, Sudan*

## Abstract

Atrial septal defects are common in patients with Ebstein malformation of the tricuspid valve and may contribute to their hemodynamic disturbances. We report our experience of trans-catheter closure of large atrial septal defects in two patients with Ebstein malformation using Amplatzer in one and Helex septal occluder in second patient with no residual shunts. There was no immediate complication, and 18 months follow up revealed clinically stable patients with normal sinus rhythm and no residual shunt. We can say that trans-catheter closure of large atrial septal defects in Ebstein malformation is feasible with a good short-term outcome. *Heart Views 2007;8(3)109–111.*

## Introduction

Ebstein malformation has been reported to represent 2% of congenital heart defects in Sudanese population which is 4 times more than the reported frequency in the Western literature<sup>1</sup>. Secundum atrial septal defect is a common finding in patients with Ebstein malformation; often leading to aggravation of the volume overload of the right atrium<sup>2,3</sup>. Secundum atrial septal defect trans-catheter closure is now considered a standard practice in patients without Ebstein malformation<sup>4,5</sup>. Since patients with Ebstein malformation have complex right atrial morphology, trans-catheter approach seems more attractive because surgical scars to this diseased right atrium might result in higher occurrence of atrial arrhythmias and postoperative complications. We report our experience with two Sudanese patients who had Ebstein malformation and large left to right shunt across secundum atrial septal defect and underwent trans-catheter closure.

## Methods and materials

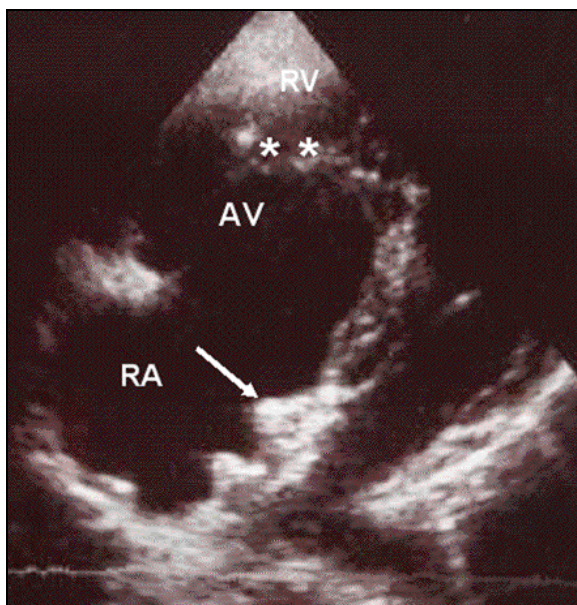
The patients were seen at the Sudan Heart Centre in 2004 and diagnosis was made by clinical and echocardiographic assessment. Ebstein malformation was diagnosed when the septal leaflet of the tricuspid valve was apically

displaced by more than 8mm per square meter of body surface area and this was referred to as displacement index. In April 2006, under general anesthesia; trans-esophageal echocardiography (TEE) and cardiac catheterization were performed and trans-catheter device closure of these atrial septal defects was attempted.

## Patient 1

A 35 year old Sudanese lady was evaluated because of history of shortness of breath with exercise for the past 2 years. Six months prior to presentation, she experienced left sided weakness involving her upper and lower limbs. This weakness resolved gradually over 3 months and eventually she regained complete motor function. Cardiac examination at presentation revealed signs of atrial septal defect. Her systemic oxygen saturation was 98% and electrocardiogram showed normal sinus rhythm, tall peak P wave, first degree atrioventricular block, RSR' pattern and no pre excitation. Trans-thoracic echocardiogram revealed Ebstein malformation of the tricuspid valve with a septal leaflet displacement index of 20, mild to moderate tricuspid regurgitation, large secundum atrial septal defect measuring 18mm with left to right shunt, dilated right

**Correspondence to:** Sulafa KM Ali, Paediatric Cardiology Department, Sudan Heart Centre, PO Box 11917, Khartoum, Sudan Phone: 00249 183 232137, Fax 00249183 232135, e-mail: sulafakhalid2000@yahoo.com.

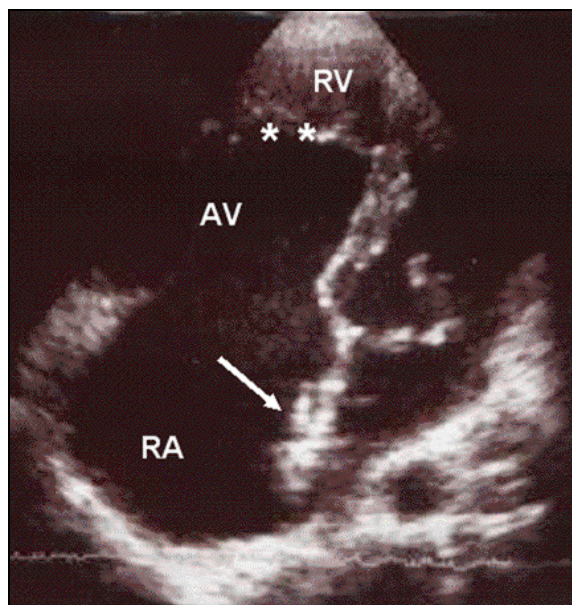


**Fig. 1:** Modified 4 chamber view of patient no. 1 showing Amplatzer device aligned with the interatrial septum (arrow), displaced tricuspid valve (stars) and atrialized right ventricle. (RA: right atrium, AV: atrialized right ventricle, RV: right ventricle).

atrium, normal left sided structures and normal left ventricle ejection fraction. The patient underwent trans-esophageal echocardiogram and cardiac catheterization, and balloon stretched atrial septal defect diameter measured 26mm. Hemodynamic assessment showed Qp:Qs of 2.6:1, mean right atrial pressure was 10, right ventricle pressure 20/0 and pulmonary artery mean pressure was 10mmHg. Under TEE, Fluoro and cine guidance, 26mm Amplatzer atrial septal defect occluder was deployed successfully with good alignment (Fig1) and no residual atrial shunt. Post procedure, right atrial pressure did not change and there were no complications. Prophylactic antibiotics and heparin were given and she was discharged 24 hours after the procedure on aspirin 100mg once a day for 6 months. On follow up at 2, 6 and 18 months, she was asymptomatic, in sinus rhythm and echocardiography showed mild tricuspid regurgitation and no residual atrial septal defect.

### Patient 2

An 8 years old female was evaluated because of a heart murmur. Physical examination showed normal growth, normal first heart sound, widely splitting second heart sound and a soft ejection systolic murmur at the pulmonary area with no clicks. Her oxygen saturation was 98% on room



**Fig. 2:** Modified 4 chamber view of patient no. 2 showing Helix device aligned with the interatrial septum (arrow), displaced tricuspid valve (stars) and atrialized right ventricle. (RA: right atrium, AV: atrialized right ventricle, RV: right ventricle).

air, electrocardiogram showed normal sinus rhythm, tall peak P wave, RSR' pattern and no pre excitation. Echocardiogram revealed Ebstein malformation of the tricuspid valve with setpal leaflet displacement index of 35, mild tricuspid regurgitation, 20mm fenestrated secundum atrial septal defect with left to right shunt, dilated right atrium, normal left sided structures and normal left ventricle ejection fraction.

Under general anesthesia, trans-esophageal echocardiogram confirmed transthoracic echo findings, and cardiac catheterization showed Qp: Qs of 2:1, mean right atrial pressure was 8mmHg, right ventricle pressure 20/0mmHg and pulmonary artery mean pressure was 10mmHg. Under TEE, Fluoro and cine guidance, 25mm Helix® (Gore) ASD occluder was deployed successfully with good alignment (Fig 2) and no residual atrial shunt. Right atrial pressure remained the same after procedure. Prophylactic antibiotics and heparin were given. During the procedure, she developed supraventricular tachycardia with a heart rate of 190 beats per minute and was aborted with Verapamil. The tachycardia recurred one hour after the procedure and was controlled by oral Amiodarone. She was discharged 48 hours after the procedure in sinus rhythm on Amiodarone and aspirin 50mg once a day. On follow up at 2, 6 and 18 months, she was clinically asymptomatic, in sinus rhythm and echocardiography showed

mild tricuspid regurgitation and no residual atrial septal defect. Amiodarone was discontinued after 6 months without recurrence of tachycardia.

## Discussion

Closure of atrial septal defect in patients with Ebstein malformation is expected to improve the right atrium volume overload. Surgical atrial septal defect closure is not without risk even in patients with otherwise normal hearts as Garson et al<sup>6</sup> reported the incidence of atrial arrhythmia in up to 12.4% of patients. Similarly, Bink-Boelkens et al<sup>7</sup> followed 204 patients who underwent surgical repair of atrial septal defect for 10 years and 18% of these patients developed atrial arrhythmia in the follow up period. Moreover, it has been reported that some cases of Ebstein's malformation deteriorated after surgical closure of atrial septal defect<sup>8</sup>.

There are few reports in the literature about percutaneous atrial septal defect closure in patients with Ebstein malformation, these reports included patent foramen ovale and small left to right shunt<sup>9,10,11</sup>. Our decision to go for interventional catheterization was based on the patient's symptoms, echocardiographic demonstration of a large left to right shunt supported by hemodynamic data obtained during cardiac catheterization. Patient # 2 developed supraventricular tachycardia during catheter manipulation in the atria, which was controlled with verapamil and then Amiodarone. Gabriella Agnoletti et al<sup>9</sup> had published an experience of 4 patients with Ebstein malformation who had atrial septal defect catheter closure and one of them needed ablation therapy after the closure. This raises concern whether all patients with Ebstein malformation should routinely have electrophysiologic studies prior to interventional treatment because of their liability to arrhythmias but this needs further studies to look at the long term results. Short term follow up showed improving symptoms in both patients and no deterioration in the tricuspid valve function.

We conclude that the less invasive transcatheter atrial septal defect closure in patients with Ebstein's malformation results in less hospital stay and good short term outcome. ?

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