

## Role of Magnetic Resonance Imaging in Evaluation of Ebstein's Anomaly

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### Abstract

**Background:** Ebstein's Anomaly (EA) is one of the uncommon Congenital Heart Disease (CHD). It accounts for about 0.5% of patients. It is defined as apical displacement of the posterior and septal leaflets of the tricuspid valve which results in atrialization of a part of the right ventricle.

**Aim of Study:** Review and discuss the role of Magnetic Resonance Imaging (MRI) findings of EA as regards the morphological and hemodynamic aspects, thus emphasize the role of MR in the assessment of myocardium as well as ventricular and valvular functions in cases of EA.

**Subjects and Methods:** Our retrospective cohort study included twenty patients who were previously diagnosed as EA at the National Heart Institute, Giza, Egypt. The clinical records and surgical details of all included patients were revised; study participants were selected from the outpatients.

**Results:** Statistical analysis revealed that Ebstein's anomaly severely affected tricuspid regurge, where 75% of patients (15 of 20) while only one patient (5%) suffered of mild regurge and four patients (20%) suffered of moderate regurge.

**Conclusion:** The diagnosis of Ebstein's anomaly is usually based on echo findings; however, Cardiac Magnetic Resonance imaging (CMR) can add useful detailed information, as well as accurate physiological evaluation for decision making in these patients. CMR in conjunction with echocardiography offers a comprehensive non invasive evaluation either for surgical management or ongoing follow-up of these patients.

**Key Words:** *Ebstein's anomaly – Congenital heart disease – Magnetic resonance imaging – Tricuspid regurge.*

### Introduction

**EBSTEIN'S** Anomaly (EA) is one of rare Congenital Heart Disease (CHD). It accounts for 0.3-0.5% of CHDs. It is defined as apical displacement of the posterior and septal leaflets of the tricuspid valve as a result of abnormal development of the Tricuspid Valve (TV) and Right Ventricle (RV) in

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early fetal life which results in RV myopathy and abnormal TV morphology that always results in variable degrees of Tricuspid Regurgitation (TR) and atrialization of a part of the right ventricle [1-4].

This anomaly has wide spectrum of clinical course depending on the degree of right ventricular myopathic affection and the degree of tricuspid displacement and regurgitation. It may result in profound Congestive Heart Failure (CHF) in early life in severe form or may remain silent throughout life; however the majority of cases present with symptoms during their life according to the degree of affection.

So most of the patients with EA usually conservatively treated and kept under close follow-up, whereas surgical intervention is indicated for those with marked RV dilation associated with progressive impairment of ventricular systolic functions [4].

The decision of surgical intervention is variable according the RV functions as it may be biventricular repair or 1.5-ventricular repair (bidirectional Glenn shunt) in case of poor right ventricular function, or it may need heart transplantation for the patients with severe left ventricular dysfunction [4]. CMRI can provide detailed and accurate information on the morphology and function of the TV and RV which are challenging by echocardiography because of the poor acoustic window. The MR findings of EA are illustrated in this study [5,6].

### Patients and Methods

A retrospective study involved twenty patients were referred to us from congenital clinic at National Heart Institute, Giza, Egypt. Our patient's age range from 10 to 40 years with mean patient

age was  $26 \pm 14$  years, 40% were  $< 18$  years. Eleven of them (55%) were female & 9 (45%) were males Fig. (6).

All gave written informed consent to participate in the study, and were examined during the period from June 2018 February 2020 in our outpatient clinic. Inclusion criteria were EA, no TV or major cardiac corrective surgery (other than atrial septal defect closure). The study protocol was approved by the ethics committee of the National Heart Institute and was in accordance with the 1975 Declaration of Helsinki.

All patients were subjected to detailed medical history, clinical examination, echocardiography, serum creatinine and CMR.

Clinical examination including measurement of trans-cutaneous oxygen saturation, blood pressure, Heart Rate (HR), body weight, and height.

CMR scans were performed on a 1.5-Tesla MRI-scanner (Siemens Medical, Erlangen, Germany) using phased array receiver coils. All patients were examined according to the protocol for EA. No sedation nor anesthesia were applied.

The protocol included localizers, stacks of Steady State Free Precession (SSFP) cine images in transversal and short-axis orientation with zero gap and field of view covering all cardiac structures and great vessels, Cine SSFP in 2 chamber (left and right), 3 chamber (left and right) and 4 chamber views. Through-plane Phase Contrast (PC) at the level of aortic valve, main pulmonary, right and left pulmonary arteries.

Consecutive short-axis views covering the LV from base to apex (TR/TE 3.4ms/1.3ms, flip angle  $50^\circ$ , FOV 300 X 340mm, matrix size 256 X 144, slice thickness 8mm), and consecutive axial cine bSSFP images covering the whole heart extending from the pulmonary bifurcation to just below the diaphragm. LGE images were acquired using a T1-weighted inversion recovery turboflash sequence 10 to 20min after bolus contrast injection (Gadopentetate dimeglumine, 0.3/kg). Measurements of ventricular volumes and functions were performed using stacks of SSFP short-axis (SAX), and transversal planes, each with full coverage of both ventricles and both atria.

End-diastolic and end-systolic areas were traced at Syngovia work-station in each slice that are stacked together to generate end systolic volume (ESV) and End-Diastolic Volume (EDV) of both atria, functional RV (fRV), atrialized RV (aRV)

and Left Ventricle (LV) Figs. (1,3), allowing calculation of Stroke Volume (SV) and Ejection Fraction (EF) of both ventricles.

Contours of the right side of the heart (RA, fRV and aRV) were performed in using the trans-axial stack plane as delineation of atrio-ventricular junction and attachments of abnormal tricuspid valve were better in this orientation Fig. (2).

Indirect quantification of the TR was done indirectly through calculation of the TV regurgitate volume by subtracting forward flow, as measured by velocity mapping of pulmonary artery (sum of both RPA & LPA), from total RV stroke volume measured on the cine images. Regurgitant fraction can then be calculated by dividing this regurgitant volume by RV stroke volume [7].

Assessment of atrial level shunt is best seen by echocardiography, but may also be visualized in cine images Fig. (1). The direction and significance of shunt were assessed by calculation the ratio between the net pulmonary flow ( $Q_p$ ) and net aortic flow ( $Q_s$ ) ( $Q_p: Q_s > 1$  denoting left to right,  $< 1$  right to left shunt).

Late Gadolinium Enhancement (LGE) sequences were taken in SAX, 2 chamber, 3 chamber and 4 chamber views to detect scar and fibrosis.

#### Statistical methods:

Data management and statistical analysis were done using SPSS vs. (IBM, Armonk, New York, United states). Numerical data was summarized as means and standard deviations. Categorical data were summarized as numbers and percentages.

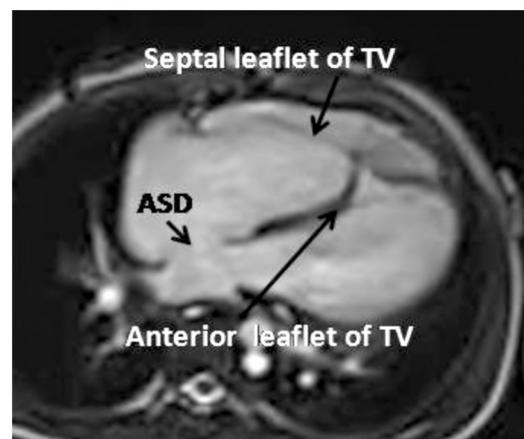


Fig. (1): MRI cine SSFP 4-chamber projection of 10 years old male patient with Ebstein anomaly showing moderate degree/grade 2 of apical displacement of the septal leaflet of the TV as well long sail like anterior leaflet and large ASD as one of common association.

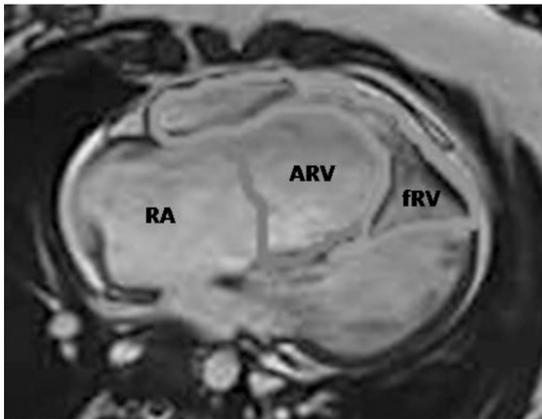


Fig. (2): MRI cine SSFP 4-chamber projection of 22 years old female patient with Ebstein anomaly showing the blue line indicates the atrialized right ventricle and red line indicates the functional right ventricle (fRV).

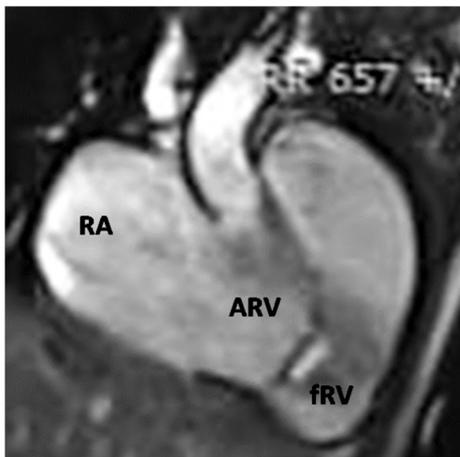


Fig. (3): MRI cine SSFP 2-chamber right projection of 34 years old male with Ebstein anomaly showing the atrialized right ventricle and the functional right ventricle (fRV).

**Results**

*Patient characteristics and severity of EA assessed by CMR are summarized:*

As regards the degree of apical displacement of TV; 5 out of 20 (25%) showed grade 1/mild displacement Fig. (4) while 11 patients (55%) showed grade 2/moderate displacement Fig. (1) and 4 (20%) showed grade 3/severe displacement Fig. (2) (Table 1) Fig. (7).

As regards the TR, most (75%) of our cases (15 of 20) showed severe TR Fig. (5), while only one patient (5%) suffered of mild TR and four patients (20%) suffered of moderate TR (Table 2) Fig. (8).

According to RV functions 55% of patients showed fair function. (EF 32% to 46%) while 45% of patients showing good RV functions >(EF >46%) (Table 3) Fig. (9).

As regards shunting; 7 patients (35%) were found to have right to left shunt while 4 patients showed left to right shunt (20%) and 9 (45%) patients were found to be negative for shunt (Tables 4, 5).

According to regional wall motion abnormality; 8 patients (40%) had abnormality of their regional wall motion but 12 patients (60%) were not affected (Table 6), Fig. (10).

Only 5% of EA patients showed impaired systolic left ventricular function while the function good in 95% of patients (Table 7), Fig. (11).

There was no pulmonary stenosis due to Ebstein anomaly since there is no cases were demonstrated in this study (as illustrated in table number one).

There were only 2 cases (10%) out of 20 patients showed RV fibrosis in late gadolenium enhancement LGE Fig. (13).

Only two cases out of 20 (10%) showed associated Mitral Regurgitation (MR) Fig. (12) (Table 8).

Females were significantly affected by Ebstein anomaly more than males ( $p=0.028$ ) using Fisher's exact test since 63% of females had regional wall motion abnormality but only 11 % of males were affected, as illustrated in table number 6; females had regional wall motion abnormality but only 11 % of males were affected, as illustrated in table number 6.

Table (1): Degree of displacement in tricuspid valve according to gender

Tricuspid valve degree of displacement	Males (n=9)		Females (n=11)		p-value
	N	%	N	%	
Grade 1	4	44.4	1	9.1	0.281
Grade 2	4	44.4	7	63.6	
Grade 3	1	11.1	3	27.3	

Fisher's exact test was used for tricuspid valve degree of displacement within the same gender and were found to be non significant ( $p=0.28$ ) either in males or females.

Table (2): Degree of tricuspid regurge according to gender.

Degree of TR	Males (n=9)		Females (n=11)		p-value
	N	%	N	%	
Mild	0	0.0	1	9.1	0.026
Moderate	4	44.4	0	0.0	
Severe	5	55.6	10	90.9	

Fisher's exact test was used to calculate the degree of tricuspid regurge according to gender. Results were significant ( $p=0.026$ ) denoting effect of Ebstein Anomaly on the tricuspid regurge.

Table (3): Right ventricular function according to gender.

Functional RV	Males (n=9)		Females (n=11)		p-value
	N	%	N	%	
Fair	3	33.3	8	72.7	0.095
Good	6	66.7	3	27.3	

Fisher's exact test was used were calculated regarding right ventricular function according to gender. Although right ventricle was found small in size but its function was not significantly affected.

Table (4): Right to left shunt according to

	Males (n=9)		Females (n=11)		p-value
	N	%	N	%	
Right to left shunt	2	22.2	5	45.5	0.5/4

Fisher's exact test was used.

Table (5): Left to right shunt according to

Left to right	Males (n=9)		Females (n=11)		p-value
	N	%	N	%	
Positive	2	22.2	2	18.2	1.0

Fisher's exact test was used.

Table (6): Regional wall motion abnormality according to gender.

Regional wall motion	Males (n=9)		Females (n=11)		p-value
	N	%	N	%	
Regional wall motion	1	11.1	7	63.6	0.028

- Females were significantly affected by Ebstein anomaly more than males ( $p=0.028$ ) using Fisher's exact test since 63% of females had regional wall motion abnormality but only 11% of males were affected.

Table (7): Left ventricular function according to

LV function	Males (n=9)		Females (n=11)		p-value
	N	%	N	%	
Fair	0	0.0	1	9.1	1.0
Good	9	100.0	10	90.9	

Fisher's exact test was used.

Table (8): Mitral regurge according to gender.

Mitral Regurge	Males (n=9)		Females (n=11)		p-value
	N	%	N	%	
Mild	1	11.1	1	9.1	1.0
No MR	8	88.9	10	90.9	

Fisher's exact test was used.

Table (9): Pulmonary stenosis in the whole study population.

Pulmonary stenosis	N (%)
Yes	0 (0.0)
No	20 (100.0)

- There was no pulmonary stenosis due to Ebsteint anomaly since there is no cases were demonstrated in this study.

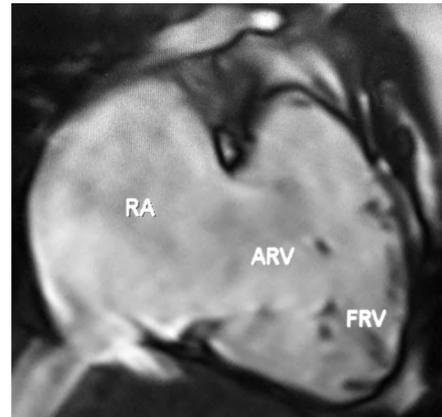


Fig. (4): MRI cine SSFP 2 chamber right of 17 years old male patient with mild form of Ebstein anomaly showing mild/grade 1 displacement of the septal leaflet of the TV with adequate volume of the functional right ventricle (fRV).

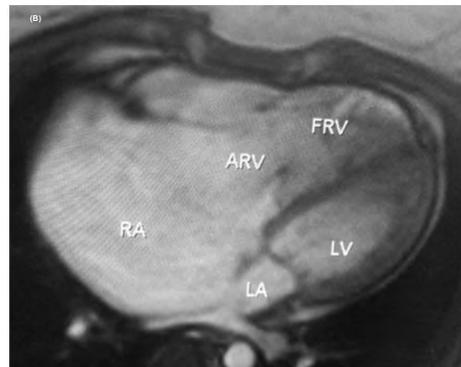
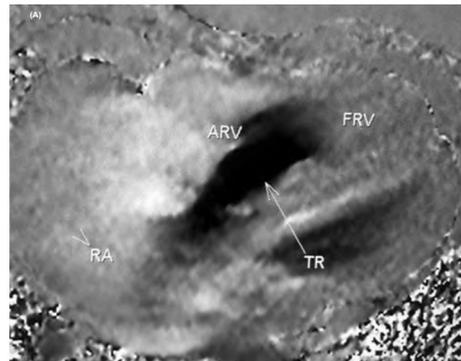


Fig. (5): (A) MRI phase contrast in plane flow sequence in 4 chamber view of 40 years old female patient showing severe TR that appear as dark jet reaching the right atrium. (B) Shows the 4 chamber view (the same anatomical plane of the phase contrast image of the same patient) showing the displaced tricuspid valve with failure of coaptation.

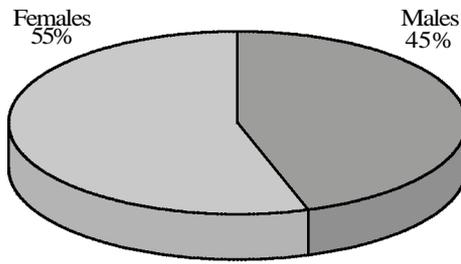


Fig. (6): Demographic characteristics in the whole study population.

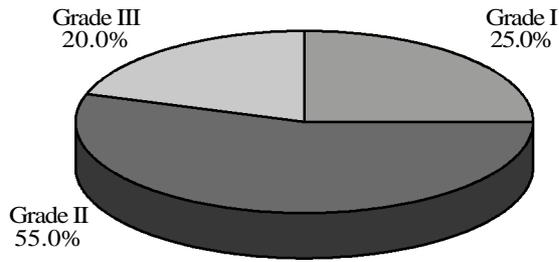


Fig. (7): Tricuspid valve degree of displacement.

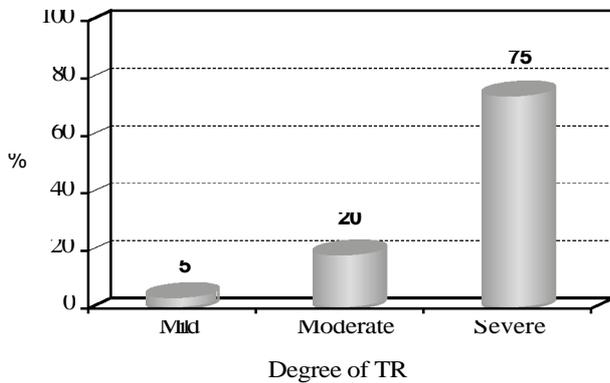


Fig. (8): Degree of tricuspid regurgitation in the whole study population.

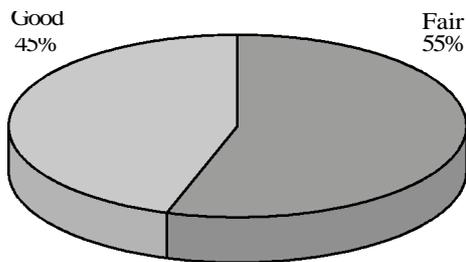


Fig. (9): Right ventricular function in the whole study population.

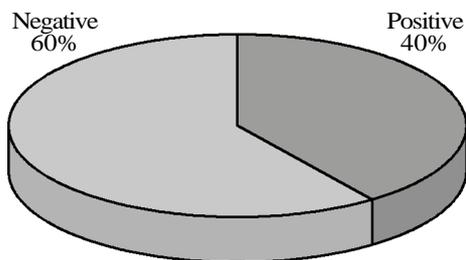


Fig. (10): Regional wall motion abnormality.

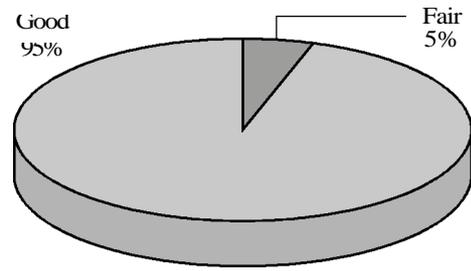


Fig. (11): Left ventricular function in the whole study population.

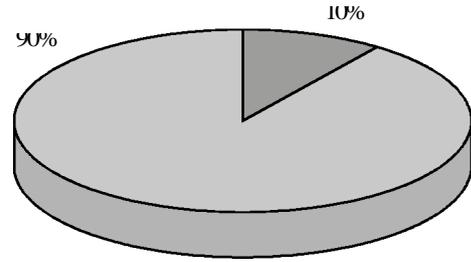


Fig. (12): Mitral regurgitation in the whole study population.

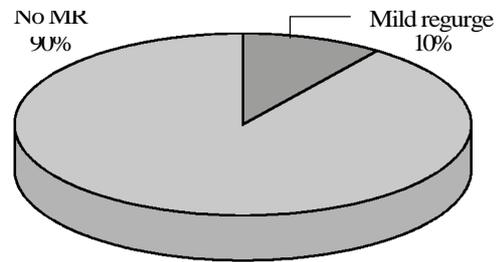


Fig. (13): Late gadolinium enhancement in the whole study population.

### Discussion

CMR allows detailed visualization of the anatomy in patients with EA. It is of great value when echocardiography image quality is inadequate due to a poor acoustic window. CMR also provides a method of accurate physiological assessment; as it is a gold standard technique for precise volumetric analysis as well as ventricular functions.

It also provide information about the direction and degree of shunt as well as degree of regurgitations.

CMR delayed contrast enhancement image is a potential tool to recognize areas of right ventricle dysplasia.

#### 1- RV and LV functional analysis:

In our study, more than half of the patients ( 55%) RV ejection fraction ranged between 32% to 46% and in the remaining 45% of patients, ejection fraction >46%, which was comparabile to Yang D. et al., 2008 [1].

*Associations of Ebstein's anomaly:*

ASD is the most common associated anomaly found in Ebstein as it is found in 55% of patients in our study (11 out of 20 patients) with 35% had right to left shunt and 20% with left to right shunt. Pulmonary stenosis was not demonstrated in our study because it is a rare finding usually found in cases with small RV associated with marked restriction in anterior leaflet movement (that was not well presented in our study).

*Late Gadolinium Enhancement (LGE) and regional motion abnormality:*

Myocardial late enhancement contrast image has the ability to precisely delineate myocardial scar associated with fibrosis. In the EA scene this is an important tool to recognize areas of dysplasia of right ventricle.

LGE was found in 2 (10%) patients. Typical LGE in Ebstein's anomaly was located in the endocardium of the septum within the Right Ventricle (RV). This low percentage may be due to the quality of patients undergone this technique which most of them are relatively of fair to good ventricular function in comparison to patients have LGE.

This proves that fibrosis is usually associated with the severity of Ebstein anomaly [8].

*Conclusion:*

The diagnosis of Ebstein's anomaly is usually based on echocardiographic findings; however, Cardiac Magnetic Resonance imaging (CMR) can add useful information enabling a detailed unrestricted visualization of cardiac abnormality, and still a gold standard method of accurate physiological evaluation without geometric assumption or ionizing radiation. CMR, in conjunction with echocardiography, offers a comprehensive non

invasive evaluation either for surgical management or ongoing follow-up of these patients.

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## دور الرنين المغناطيسي للقلب فى تقييم شذوذ ابشتاين كعيب خلقى للقلب

شذوذ ابشتاين هو نزوح خلقى غير شائع للنشرات الخلفية والحاجزية لصمام ثلاثى الشرفات باتجاه قمة البطن الأيمن. هذه النتائج فى إحلال جزء من البطن الأيمن، دائماً ما يكون هناك قصور ثلاثى الشرف.

أكثر من ٩٠٪ من حديثى الولادة يعانون من زرفة ثانوية إلى تحويلة من اليمين إلى اليسار على مستوى الأذين أو فى وجود تضيق صمام رئوى أو رتق.

متلازمة إيبيستينشن يضم ٠.٥-٠.٠٪ من أمراض القلب الخلقية.

الشذوذ التنموى لصمام ثلاثى الشرف الذى يحتوى على مجموعة واسعة من التشريح المرضى. مع تطور التقنيات الجراحية المختلفة، فإن التقييم الشامل للسماط المورفولوجية ونوع شذوذ إبشتاين يفترض أهمية عملية أكبر. على الرغم من أن تشخيص شذوذ إيبيستين عادة ما يكون مستقيماً ويعتمد على نتائج تخطيط صدى القلب، إلا أن هناك قيوداً فى تحديد التشريح المرضى فى بعض الحالات. الغرض من هذا المقال هو توضيح النتائج الرنين المغناطيسى فى أنواع مختلفة من متلازمة إبشتاين، فضلاً عن درجة القصور ثلاثى الشرف والحق فى التنقل إلى اليسار. وقد أتبع العلاج الجراحى لشذوذ إبشتاين مسارين متوازيين: الإستبدال وإعادة بناء البلاستيك من صمام ثلاثى الشرف. بما أن الطريقة الجراحية مصممة لكل مريض، فإن التقييم الدقيق للسماط المورفولوجية ونوع شذوذ إبشتاين أمر حاسم. تصنيف هذه الحالة الشاذة وفقاً لنظام Carpentier et al. مفيد جداً لهذا الغرض. إن إعادة بناء الصمام ثلاثى الشرف بإستخدام حلقة إصطناعية وفتحة عمودية من الأذين اللىلى والحلقة العصبية الأذينية، مع (النوعين ب و س) بدون (نوع أ) من تقطيع البطن الأذينى قد حسنت النتائج الجراحية. يمكن تصوير القلب بالرنين المغناطيسى توفير معلومات دقيقة عن مورفولوجيا وحالة كل الهياكل داخل القلب وخارج القلب بسبب مزاياه الكامنة. يتم توضيح النتائج الرنين المغناطيسى من أنواع مختلفة من شذوذ ابشتاين فى هذه الرسالة المصورة.

الهدف من هذه الدراسة: هو تقييم دور التصوير بالرنين المغناطيسى فى تشخيص أنواع مختلفة من إيبيستين الشاذة والكشف عن إرتباطاته المشتركة وكذلك تقييم المرضى سيخضعون لإصلاح جراحى.