**Original Article**

**Intraoperative Transesophageal Echocardiography in the Operation of Ebstein’s Anomaly: A Retrospective Study**

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**Abstract**

**Background:** Ebstein’s anomaly (EA) has various spectrums in clinical and anatomic features. This study aimed to report the experience of two-dimensional intraoperative transesophageal echocardiography (2D-ITEE) during the EA surgery and to analyze the characteristics of the tricuspid valve (TV) by comparing the data from 2D-ITEE with the results from the surgery.

**Methods:** 2D-ITEE data of 164 patients with EA who were operated in the First Hospital of Tsinghua University between July 2004 and April 2014 were retrospectively analyzed in this study. 2D-ITEE was applied in all patients. Downward displacement distances were measured, and the numbers of downward displacement or absent leaflets were compared with that of the surgery and with that of the two-dimensional-transthoracic echocardiogram (2D-TTE). Data comparison was performed using the Chi-square test.

**Results:** The anterior leaflet partial or total downward displacement was 37.76 ± 17.50 mm in 54 cases, absent in one patient; septal leaflet downward displacement was 29.07 ± 12.34 mm in 134 cases, absent in 17 cases; and posterior leaflet downward displacement was 43.18 ± 19.16 mm in 115 cases, absent in 34 cases. Statistically, there was no significant difference between the results from 2D-ITEE and that of 2D-TTE. The consistency rates of 2D-ITEE with operation for septal and posterior leaflets were 93.2% and 96.1%, respectively, while the rate for anterior was only 40.1%, which was significantly different. Color Doppler flow image showed severe regurgitation in 150 cases and moderate in 14 cases. After surgical correction, moderate regurgitation of TV was found in 58 cases with 2D-ITEE, repair was performed again until the effect was satisfied. No complication occurred relating to the use of 2D-ITEE.

**Conclusions:** 2D-ITEE could help diagnose anterior leaflet, evaluate the effect of TV repair, increase operational success rate, and reduce complication.

**Key words:** Ebstein’s Anomaly; Echocardiography; Two-dimensional-transesophageal Echocardiography

**INTRODUCTION**

Ebstein’s anomaly (EA) is a rare complex congenital heart disease, occurring in about 1–5/200,000 births and accounting for <1% of all cases of congenital heart diseases.[1] This rare congenital malformation was first described by Ebstein in 1866,[2] and the first echocardiographic diagnosis of EA was reported by Lundstrom in 1969.[3] As we know, the spectrum of clinical and anatomic features of EA is wide and each patient has his/her own different characteristics.[4]

The pathological characteristics mainly include dilation of the tricuspid annulus, downward displacement of the leaflets, atrialized portion of the right ventricle (RV), and malformation of the anterior leaflet, including redundancy, fenestrations, and tethering.[5,6] Most articles reported that anterior leaflet rarely displaced from the tricuspid annulus.[7-9]

Echocardiography is currently the main diagnostic choice for EA and free from the need for cardiac catheterization.[10] The preferred treatment for EA is tricuspid repair other than valve replacement because it has better effect and avoids long-time anti-coagulation medicine. As a reliable tool, two-dimensional intraoperative transesophageal echocardiography (2D-ITEE) has been routinely used for establishing intraoperative diagnosis, monitoring hemodynamic changes, and evaluating the effect of tricuspid...
repair. However, there was a scarce in literature reporting 2D-ITEE data of large series of EA. In this report, we focused on the experience of 2D-ITEE in the repair operation of EA.

**Methods**

**Ethical approval**

As a retrospective study and data analysis was performed anonymously, this study was exempt from the ethical approval and informed consent from patients.

**Patients**

From July 2004 to April 2014, 164 EA patients, who underwent surgical treatment in the First Hospital of Tsinghua University (China), including 69 males and 95 females, between the age of 5 months and 63 years (median: 20.6 ± 15.6 years), were retrospectively reviewed. Preoperative electrocardiogram, chest X-ray, and two-dimensional-transthoracic echocardiogram (2D-TTE) were routinely performed. 2D-TTE was used to evaluate intracardiac anatomy, left ventricular function, and motion and regurgitation of tricuspid valve (TV). The indications for surgery treatment included one or more of the followings: dyspnea, progressive exercise intolerance, heart failure (New York Heart Association functional Class III or IV), tachyarrhythmia (not controllable by medication or amenable to catheter-based intervention), and significant associated lesions. Progressive cardiomegaly and higher cardiothoracic ratio (>0.65) were also additional indications for surgery. During the repair operation, 2D-ITEE was applied in all patients. Associated anomalies were listed in Table 1.

**Instruments and inspection section**

All 2D-ITEE echocardiograms were performed with GE VIVID I Cardiac Ultrasound system (General Electric Company, USA) equipped with two multi-plane TEE probes. The 9T-RS probe was usually used in patients weighing <15 kg and the 6T-RS probe for in patients weighing over 15 kg. After general anesthesia, the 2D-ITEE probe was inserted with the help of a laryngoscope. Before operation, 2D-ITEE was used to detect the morphology, size, and motion of three tricuspid leaflets and both ventricles’ function. According to the 2D-ITEE guidelines published by the American Society of Anesthesiologists and the Society of Cardiovascular Anesthesiologists,[11] the distance of downward displacement of TV was measured using a caliper tool. The reference point was the root of anterior mitral valve leaflet for septal leaflet, coronary sinus (CS) for posterior leaflet, and normal tricuspid annular for anterior leaflet. The distance from reference point to the actual position of tricuspid leaflet was collected from mid-esophageal 4-chamber view, RV inflow-outflow view, CS view, and etc. Mid-esophageal 4-chamber view clearly showed atrial septum, ventricular septum, mitral valve, TV, chordae, and papillary muscle. The dimension and function of the RV, right atrium, and atrialized RV should also be evaluated. The motion and position of anterior and septal leaflets were shown clearly, and the downward distance was measured. Color Doppler was applied to assess the location and grade of tricuspid regurgitation.

Mid-esophageal RV inflow-outflow view showed the position of anterior and septal leaflets and supplies more views and information of the anterior leaflet. In this view, the posterior leaflet of the TV was on the left side of the display and the anterior leaflet on the right side.

The CS view was obtained by tilting back the probe on the basis of 4-chamber heart view, exposing the ostial of the CS. This view showed the position and the downward displacement degree of tricuspid anterior and posterior leaflet, especially the posterior leaflet. The downward distance was measured.

Color Doppler was combined in all views to evaluate the regurgitation of TV [Figure 1]. These views are also crucial after valvuloplasty in evaluating the function and motion of TV, especially important in identifying possible stenosis and regurgitation of TV after repair. In some cases, extra-large anterior leaflet tissue or autograft pericardium was used to rebuild leaflet in hypoplasia or absent septal leaflet.

**Statistical analysis**

All analyses were performed with SPSS software for Windows version 20.0 (SPSS, Inc., Chicago, IL, USA). Data are expressed as mean ± standard deviation (SD). Data

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**Table 1: Concomitant cardiac malformation of patients with Ebstein’s anomaly**

| Lesion       | Number of patients |
|--------------|--------------------|
| ASD          | 60                 |
| PFO          | 30                 |
| VSD          | 4                  |
| PDA          | 4                  |
| LSVC         | 2                  |
| PECVD        | 1                  |
| CTGA         | 1                  |

ASD: Atrial septal defect; PFO: Patent foramen ovale; VSD: Ventricular septal defect; PDA: Patent ductus arteriosus; LSVC: Left superior vena cava; PECVD: Partial endocardial cushion defect; CTGA: Corrected transition of great artery.

**Figure 1:** The aortic short-axis view shows severe tricuspid regurgitation in color Doppler mode. RA: Right atrium; RV: Right ventricle.
comparison was performed using Chi-square test. A $P < 0.05$ was considered statistically significant.

**RESULTS**

**Data of downward displacement and absent leaflets in two-dimensional intraoperative transesophageal echocardiography**

The results from 2D-ITEE were similar with the data from 2D-TTE and had no statistically significant difference [Table 2]. The anterior leaflet was detected to be redundant as sail shaped in 109 cases, partial or total downward displaced in 54 cases, and almost absent in one case, the mean downward distance was $37.76 \pm 17.50$ mm. The anterior leaflet was pulled by abnormal chordate tendineae; the range of its motion was significantly decreased. The septal leaflet was detected obviously downward displaced in 134 cases and absent in 17 cases, the average downward displacement was $29.07 \pm 12.34$ mm. Most septal leaflets were found to be dysplasia, short, stiff, decreased motion, and poorly coapted with other leaflets in systole. 2D-ITEE showed posterior leaflet downward displacement in 115 patients, with the average distance of $43.18 \pm 19.16$ mm; in 34 patients, posterior leaflet was absent. The posterior leaflet was found abnormally developed, thickened, and short [Table 3].

**Comparison between data from two-dimensional intraoperative transesophageal echocardiography and surgery**

However, the results obtained during operation were not exactly the same compared to that of 2D-ITEE. The number of total and partial downward displacement of anterior leaflet counted up to 133 cases and absent in four cases, which is much higher than that by 2D-ITEE. The consistency rate of diagnosis between 2D-ITEE and operation was only 40.1%. The final operation results showed that the morbidity of anterior leaflet was 83.5%, and it has significant difference compared with that of the 2D-ITEE [Figure 2]. The septal leaflet with downward displacement counted up to 126, and 36 was absent from the operation. The consistency rate in diagnosis using 2D-ITEE and operation was 93.2%. While the number of downward displaced posterior leaflet was 114, up to 41 from the operation was absent, and the consistency rate of diagnosis is 96.1% between the ITEE and operation results.

**Table 2: Number of downward and absent tricuspid leaflets among patients with Ebstein’s anomaly examined by 2D-ITEE and 2D-TTE**

| Leaflets | Number of the downward patients | Number of the absent cases |
|----------|---------------------------------|---------------------------|
|          | 2D-ITEE | 2D-TTE | $\chi^2$ | $P$ | 2D-ITEE | 2D-TTE | $\chi^2$ | $P$ |
| ATL      | 54      | 52     | 0.056    | 0.81 | 1       | 1       |          |     |
| STL      | 134     | 132    | 0.080    | 0.78 | 17      | 16      | 0.034    | 0.85 |
| PTL      | 115     | 110    | 0.354    | 0.55 | 34      | 32      | 0.076    | 0.78 |

ATL: Anterior tricuspid leaflet; STL: Septal tricuspid leaflet; PTL: Posterior tricuspid leaflet; 2D-ITEE: Two-dimensional intraoperative transesophageal echocardiography; 2D-TTE: Two-dimensional transthoracic echocardiogram.

**Table 3: Number and downward distance among patients with Ebstein’s anomaly examined by 2D-ITEE**

| Leaflets | Number of patients | Downward distance (mm) | Absent cases |
|----------|--------------------|------------------------|--------------|
| ATL      | 54                 | 37.76 ± 17.50          | 1            |
| STL      | 134                | 29.07 ± 12.34          | 17           |
| PTL      | 115                | 43.18 ± 19.16          | 34           |

ATL: Anterior tricuspid leaflet; STL: Septal tricuspid leaflet; PTL: Posterior tricuspid leaflet.

**Value of two-dimensional intraoperative transesophageal echocardiography during the operation**

After surgical correction, moderate regurgitation was still detected in 58 cases by 2D-ITEE. Moreover, regurgitation was mild or disappeared following a short return to cardiopulmonary bypass and revising of TV plasty.

**DISCUSSION**

In this study, the data of 2D-ITEE were analyzed including the downward displacement distance, the number of downward displacement, and absent leaflet. There was no difference between the result from 2D-ITEE and that of 2D-TTE and also posterior and septal leaflet between the data from 2D-ITEE and those from surgery. However, the downward displacement in anterior leaflet was not consistent between the surgery and 2D-ITEE.
2D-ITEE can offer better images compared with conventional 2D-TTE. As the probe is located in the esophagus, imaging scanning does not interfere with operation. Furthermore, the probe can remain in a relatively fixed position for longer time, allowing successively contrast observation of cardiac structural changes. 2D-ITEE can show valvular development, leaflet motion, displacement, and whether there is perforation or not. In addition, 2D-ITEE can clearly show other associated cardiac abnormalities. During the operation, 2D-ITEE can provide continuous monitoring and assessment of regional and overall cardiac function so as to discover the latest changes of the heart.[12] At the end of the operation, 2D-ITEE could evaluate the operation effect, so remedial measures could be carried out if there is any problem. 2D-ITEE can give postoperative assessment of hemodynamic changes to guide treatment optimization.[13] After cardiopulmonary bypass, 2D-ITEE would demonstrate valvuloplasty effect and whether there is residual leakage.[14] Since ITEE can provide a television image, it has become an essential auxiliary device for EA surgery.[15] EA refers to a condition with downward displacement and hypoplasia of the tricuspid leaflets, annular dilation, valvular insufficiency, formation of atrialized RV, and might be associated with other anomalies.[16] Carpentier et al.17 had proposed the classification of EA in four types. However, our operation results showed that the number of the downward displacement, hypoplasia, and absent anterior leaflets hold a great proportion, and it counted up to 83.5%. The anterior leaflet downward displacement by 2D-TTE was often overlooked, maybe because most of the anterior leaflet downward displacements occurred partially, not the whole leaflet displaced like that of the posterior or septal leaflet. Through respective analysis of the data, the position of anterior leaflet downward displacement was discovered to occur at the junction between the anterior and posterior leaflets, which is quite difficult to observe. In our experience, usually, the major part of the anterior leaflet was not downward displaced and attached in the normal position of the tricuspid annulus, while, often with poor leaflet development, reduced motion, poor development of the papillary muscles and chordae tendineae. Abnormal pulling to papillary muscle could give some information about the existing anterior leaflet downward displacement. The septal leaflet is usually often involved, significantly hypoplastic, spirally downward, and also absent leaflet can exist. The postleaflet is also often affected, it can be malformed or just a remnant, and might also be absent. The RV is usually divided into two parts by the downward displaced leaflets, forming the atrialized RV between the normal annulus and downward displaced leaflets. The range of atrialized RV is related to the severity of the lesion.

The limitation of this study mainly lied in the lack of quantitative data in the facet of the anterior leaflet downward displacement or dysplasia. In conclusion, routine use of 2D-ITEE in cardiac surgery could revise the diagnosis before the cardiopulmonary bypass, evaluate surgical effect after bypass operation, reduce operation complications, and avoid reoperation, thus to make the heart operation safer.

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**Conflicts of interest**
There are no conflicts of interest.

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