ECHOCARDIOGRAPHIC DIAGNOSIS OF EBSTEIN ANOMALY

Abstract. This study examined the value of echocardiography in the diagnosis of Ebstein anomaly using data collected from 12 patients with Ebstein anomaly. These patients were hospitalized between March 2015 – October 2019 in the department of paediatric cardiac surgery in Timofei Moșneaga Republican Clinical Hospital, Chisinau Moldova. Their pre-operative echocardiography description of the tricuspid valve and the description of the surgeon from the surgery were compared. Results showed that the data obtained by echocardiography were very similar to the intraoperative findings that were reported by the surgeon in most patients.

Keywords: Ebstein anomaly, echocardiography, tricuspid regurgitation, atrialized right ventricle, congenital heart disease

ABBREVIATIONS:
ASD – atrial septal defect
VSD – ventricular septal defect

INTRODUCTION:
Ebstein anomaly is a rare congenital heart disorder occurring in ≈1 per 200 000 live births and accounting for <1% of all cases of congenital heart disease [1].

It is a downward displacement of an abnormal tricuspid valve into the right ventricle.

The defect arises from a failure of the normal process of delamination by which the tricuspid valve is separated from the right ventricle's myocardium (Fig. 1) [2].
Fig. 1: Tricuspid valve delamination [3]

The right ventricle is thus divided into 2 parts by the abnormal tricuspid valve: The first, thin walled "atrialized" portion, is continues with the cavity of the right atrium. The second, often smaller portion is consists of normal ventricular myocardium (Fig. 2) [2].

Fig. 2: Severe Ebstein’s malformation of the tricuspid valve (4 chamber view) [1,4]

The right atrium is enlarged due to tricuspid regurgitation, although the degree is extremely variable.
Ebstein’s anomaly showing a grossly displaced septal leaflet (arrow).

The anterior leaflet is severely tethered and nearly immobile. The functional right ventricle (RV) is small. ARV [1]

In more severe forms of Ebstein anomaly the effective output from the right side of the heart is decreased due to combination of the poorly functioning small right ventricle, tricuspid valve regurgitation, and obstruction of the right ventricle by the large sail-like anterior tricuspid valve leaflet [2].

Ebstein anomaly is the only congenital heart lesion that has a range of clinical presentations, from the severely symptomatic neonate to an asymptomatic adult [5].

The prognosis varies with the severity of the disease, although an early accurate diagnosis, as well as the advances in diagnostic and surgical techniques and postoperative care has led to improvements in outcome.

Timely diagnosis of a patient with Ebstein anomaly – will allow to treat and operate (if needed) in time- preventing possible irreversible heart damage and different complications, which in turn will improve the clinical condition of the patient and his prognosis.

Aim: To evaluate the value of echocardiography in the diagnosis of Ebstein anomaly.

Objective: Compare the echocardiography reports, with intraoperative findings reported by the surgeon.

MATERIALS AND METHODS:

In this study participated 12 patients with Ebstein anomaly who were hospitalized between March 2015 – to October 2019 in the department of paediatric
cardiac surgery (of congenital heart diseases) in Timofei Moșneaga Republican Clinical Hospital, Chisinau Moldova.

Among the 12 patients - 6 were males, and 6 females, between the ages 2 – 58 years.

The patients were evaluated by echocardiography. Using echocardiography the following parameters were evaluated:

– Diameter of the tricuspid valve’s fibrous ring
– The grade of tricuspid regurgitation
– The morphology of the tricuspid valve and its cusps
– Displacement of the tricuspid valve’s cusps towards the apex
– Presence / absence of septal defects (ASD / VSD)

These parameters from the echocardiography findings were compared to the operative reports that were written by the surgeon (from those who had surgery during the current hospitalization).

Among the 12 patients from the study – 9 patients had surgery:

4 of them underwent tricuspid valve repair, 4 had tricuspid valve replacement, and 1 had Glenn procedure (bi-directional cavopulmonary anastomosis).

Among the 9 patients who were operated – 7 of them were operated during the current hospitalization, and 2 of them were operated in the past – before the current hospitalization in Timofei Moșneaga Republican Clinical Hospital.

OWN RESULTS AND DISCUSSION

General data about the patients included in the study:

General data about the patients are presented in Table 1:

Table 1

| Patient nr. | Age | Gender | Diagnosis | Had corrective surgery in the past | Type in Carpentier classification | Tricuspid regurgitation | Septal defect (ASD/VSD) | Others | NYHA class |
|-------------|-----|--------|-----------|-----------------------------------|-----------------------------------|------------------------|------------------------|--------|------------|
| 1           | 2   | female | Ebstein anomaly | - | type C | grade 3-4 | ASD | Severe pulmonary hypertension | |
| No | Sex | Age | Diagnosis                          | Procedure/Event Description                                                                 | Apical muscular VSD (possibly large) | WPW syndrome | Pulmonary artery stenosis | Pulmonary hypertension | AV block | Arterial hypertension | Infectious endocarditis (with tricuspid vegetation) | LAD stenosis (75%) | Pericardial effusion | Possibly ASD (was not excluded) | Moderate pulmonary valve stenosis |
|----|-----|-----|-----------------------------------|-----------------------------------------------------------------------------------------------|-------------------------------------|---------------|--------------------------|------------------------|----------|------------------------|---------------------------------------------------|---------------------|-----------------------------|--------------------------------|----------------------------------|
| 2  | male| 9   | Ebstein anomaly                   |                                                                                                | -                                   | Type B        | grade 4                  | -                      | -                    | class 3                             |                                                                  |                    |                             |                                |                                 |
| 3  | male| 17  | Ebstein anomaly                   |                                                                                                | -                                   | -             | grade 4                  | ASD                    | Pulmonary hypertension | class 3                             |                                                                  |                    |                             |                                |                                 |
| 4  | male| 18  | Ebstein anomaly                   | After tricuspid valvuloplasty – 2 months before (came due to pleural effusion)               | -                                   | grade 2-3     | -                        | -                      | -                    | class 2                             |                                                                  |                    |                             |                                |                                 |
| 5  | male| 20  | Ebstein anomaly                   | After Glenn procedure (bi-directional cavopulmonary anastomosis) and repair of ASD 15 years before | -                                   | grade 2       | -                        | -                      | -                    | class 2                             |                                                                  |                    |                             |                                |                                 |
| 6  | male| 31  | Ebstein anomaly                   |                                                                                                | -                                   | type C or D   | grade 3                  | VSD                    | WPW                  | supraventricular extrasystole |                                                                  |                    |                             |                                |                                 |
| 7  | female| 31 | Ebstein anomaly                   |                                                                                                | -                                   | type C        | grade 3                  | ASD                    | Pulmonary hypertension | class 2                             |                                                                  |                    |                             |                                |                                 |
| 8  | female| 44 | Ebstein anomaly                   |                                                                                                | -                                   | type A        | grade 2-3                | -                      | Arterial hypertension – grade 2 | class 3                             |                                                                  |                    |                             |                                |                                 |
| 9  | female| 45 | Ebstein anomaly                   |                                                                                                | -                                   | -             | grade 4                  | -                      | LAD stenosis (75%) | class 3                             | Infectious endocarditis (with tricuspid vegetation) |                    |                             |                                |                                 |
| 10 | male| 51  | Ebstein anomaly                   |                                                                                                | -                                   | -             | grade 3-4                | ASD                    | Pulmonary hypertension | class 3                             |                                                                  |                    |                             |                                |                                 |
| 11 | female| 54 | Ebstein anomaly                   |                                                                                                | -                                   | type C        | grade 4                  | VSD                    | Pericardial effusion |                                                      |                                                                  |                    |                             |                                |                                 |
| 12 | female| 58 | Ebstein anomaly                   |                                                                                                | -                                   | -             | grade 2-3                | Possibly ASD (was not excluded) | Moderate pulmonary valve stenosis |                                                      |                                                                  |                    |                             |                                |                                 |
Comparison of description of the tricuspid valve from the pre-operative echocardiography, and the description in the operative report:

In order to evaluate the value of echocardiography in the diagnosis of Ebstein anomaly and to assess its ability to accurately determine the morphology of the tricuspid valve, the pre-operative echocardiography description and the description from the operative reports were compared.

The comparison between the data about the morphology of the tricuspid valve from the echocardiography reports, with the intraoperative findings reported by the surgeon, is presented in table 2:

**Table 2**

| Patient no. | Age | Gender | ASD/VSD | Tricuspid fibrous ring | Cusps morphology | Displacement towards the apex |
|-------------|-----|--------|---------|------------------------|------------------|-------------------------------|
| 1           | 2   | Female | VSD 8-10 mm | ASD – 10 mm | Was not mentioned | Septal cusp with fixation to the apex |
|             |     |        | no VSD   |           |                  | 50 mm Septal cusp |
| 2           | 9   | Male   | Patent foramen ovale | 29 mm Dilated - 41 mm | The septal cusp is attached to the septum and has limited movement | Retracted, immobile septal cusp |
| 3           | 17  | Male   |          | 19-31 mm Dilated | Coaptation defect of 7-8 mm | - |
| 4           | 18  | Male   |          |           |                  | - |
| 5           | 20  | Male   |          |           |                  | - |
| 6           | 31  | Male   |          |           |                  | - |
| 7           | 31  | Female | ASD – ostium secundum 7 mm | ASD – ostium secundum 12 mm | Was not mentioned | Septal and posterior cusps are displaced towards the septal cusp |
|             |     |        |         |           |                  | 30-35 mm 25 mm |
|             |     |        |         |           |                  | mm mm |
### Table continuation 1

| No | Age | Sex | ASD | ASD | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Valve | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cusp | Cus...
The preoperative echocardiography results of 72% of them were compatible – with almost the same findings as was described later by the surgeon in the operative report. (5/7)

However, the preoperative echocardiography results of 28% of them (2/7) have shown to be significantly different from the operative report that was described by the surgeon.

In patient number 1 – the echocardiography results describe a displacement of the septal cusp by 50 mm, whereas the operative report describes a displacement of the septal and anterior cusp by only 10 mm.

In patient number 9 – there is also a difference between the preoperative echocardiography and the operative report. The surgeon described a rupture of the anterior cusp’s cords, which was not mentioned in the preoperative echocardiography report.

**CONCLUSION:**

The data obtained by echocardiography are compatible, and very similar to the intraoperative findings that were reported by the surgeon.

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