Long term follow-up of fetal cases with tricuspid valve anomalies


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INTRODUCTION

Severe tricuspid valve anomalies (Ebstein and Non-Ebstein type) are known to be frequently complicated in utero by hydrods fetalis (FH) or arrhythmias and may have poor outcome already in utero or after birth, due to heart failure. Surgical correction is also associated with a high mortality. Long term outcome of survived cases is not still under question.

OBJECTIVES: to analyse retrospectively the characteristics and long term outcome of our cases with tricuspid valve anomaly diagnosed at fetal echocardiography.

MATERIALS AND METHODS

Between 1986 and June 2012 41 fetuses were diagnosed in our center to have Ebstein (Eb) or non-Ebstein dysplasia (NEb) of the tricuspid valve, at 21 to 36 weeks of gestation (wg), median 29:

19 - Ebstein anomaly
22 - Non-Ebstein dysplasia, 1 with associated mitral dysplasia and 2 with secundum atrial septation

2 Ebst had associated extracardiac anomalies:
- 1 labialpalatoschisis, 1 spina bifida + mielomeningocele
3 Ebst had familial history of congenital heart disease/extracard. anomalies:
- one had a parent with TGA and a previous suhbling with Prune-Belly s.
- the father of another one had Robertsonian translocation;
- the mother of the 3rd one had subvalvular aortic stenosis

2 women was on lithium an another one on gardenal in pregnancy. Another woman was taking lithium for 10 years till the current pregnancy.

Echocardiographic features, course in utero and after birth were analysed (median follow-up 5 yrs 6m-27yrs).

STATISTICAL ANALYSIS: Wilcoxon and Chi-square tests in cases who died and in survivors for variables: grade of tricuspid regurgitation (TR), Celerierman index (CInd), cardiothoracic ratio (CR), fetal hydrods (FH), pulmonary stenosis/atresia (PS, PAtr).

RESULTS

ECHOCARDIOGRAPHIC FEATURES:
------13/19 fetuses with Ebstein had a moderate-severe displacement of the TV and severe TR at presentation or worsened in utero.
3 had moderate pulmonary stenosis (PS) and 5 pulmonary atresia (PAtr); the case with lithium therapy in pregnancy was of a mild entity.
------13/22 cases of NEbstein had severe TR, others mild-moderate; 5 had moderate organic PS and 5 functional-organic pulmonary atresia.
------Fetal hydrods (FH) was present at presentation at 29-31 wg in 7/41 cases (17%)
- 5 Ebst, 2 NonEbstein

OUTCOME

5 infants with Ebstein and 6 with NEbstein were OPERATED . 4 NEbstein survived, 1 being reoperated after 1 year;
The case with associated mitral dysplasia improved the TR but required a trivalve plasty and is alive at 8 yrs.

Out of 13 cases that died, 5 had FH, 2 severe arrhythmias, all a higher grade of TR, CInd>1, CR>0.65 and 9 had PAtr.
Another infant died at 3 years due to TACHYARRHYTHMIAS and severe worsening of TR.

TOTAL MORTALITY was 13/29 = 44.8% cases that continued pregnancy ,
4/8 with FH, 9/16 Ebstein (50%), 4/16 NEbstein = 25%

CONCLUSIONS

Anatomic features were determinant for the outcome:
1/ the degree of displacement of TV and entity of TR in cases of Ebstein and entity of TR in Non-Ebstein cases
2/ Association of functional or organic pulmonary stenosis/atresia
Severe supraventricular arrhythmias led to IUD in 1 and contributed to the worsening of the clinical situation and death at 3 years in the 2nd one (presented numerous abnormal pathways at electrophys.study).

The variables TR, CInd and PAtr of dead cases were highly significantly different with respect to the survivors (p<0.002-0.006).

21 cases with milder forms are alive at 6m-27 yrs, stable or improved (5 Ebst, 16 NEbst).

<table>
<thead>
<tr>
<th>Table 1: Fetal Hydrods - 7 cases</th>
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<tbody>
<tr>
<td><strong>Type of anomaly</strong></td>
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<tr>
<td>---------------------</td>
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<tr>
<td>5 Ebstein</td>
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<td>2 Non Ebstein</td>
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<th>Table 2: Characteristics and outcome of operated cases</th>
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<tbody>
<tr>
<td><strong>Type of anomaly</strong></td>
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<tr>
<td>---------------------</td>
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<tr>
<td>5 Ebstein</td>
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<tr>
<td>5 Non Ebstein</td>
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<tr>
<td>1 Non Ebst +MV dyspl.</td>
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<tr>
<th>Table 3: Echocardiography</th>
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<tr>
<td>Ebstein,31 w.g., died of SVT</td>
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<tr>
<td>Non-Ebstein + PS Died after birth</td>
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<tr>
<td>Tric.+mitral dysplasia, operated, alive</td>
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Comments

Our data confirm a variable spectrum of tricuspid valve anomalies presenting in utero.
Severe variants with heart failure or arrhythmia have a poor outcome, milder forms of both variants may stabilize after birth, mainly the cases with Non-Ebstein dysplasia.