Quantitative Anatomy of Taussig-Bing Anomaly

Leo A Bockeria¹, Osman A Makhachev¹,², Margarita V Gorgeeva¹, Marina S Panova¹, Tatiana Yu Philippkina¹, Titalav Kh Khiriev¹ and Sergey B Zaets¹

¹Bakoulev Center for Cardiovascular Surgery, Moscow, Russia
²Dagestan Center for Cardiology and Cardiovascular surgery, Makhachkala, Russia

Abstract

Background: Taussig-Bing is one of the variants of the double outlet right ventricle. The characteristic feature of the Taussig-Bing anomaly (TBA) that makes it different from partial transposition of the great arteries is sub pulmonary ventricular septal defect. Morphometric studies of hearts with TBA are limited and in the majority of cases do not contain quantitative assessment of intracardiac structures. The aim of this study was to measure intracardiac structures (e.g. cardiac mass, diameter of valves, thickness of the myocardium), to assess their deviation from the individual norm, as well to determine how these parameters are influenced by anatomical variant of the lesion, anthropometric variability or pulmonary hypertension.

Methods: Thirty pediatric hearts with TBA underwent morphometric assessment. The mean age at the moment of death was 0.4 years. All patients had pulmonary hypertension of grade I-II (n=24) or grade III-IV (n=6) per Heath-Edwards. Morphometric assessment included the measurement of the following parameters: cardiac mass, diameter of the fibrous annulus of all cardiac valves (mitral, tricuspid, aortic, and pulmonary), myocardial thickness of the both right and left ventricle. Results of valvarul morphometry were compared with individual normative data presented by Schulz DM and Giordano DA, and Z-score index was calculated.

Results: Cardiac mass, right ventricular myocardial thickness and the diameter of pulmonary valve significantly exceeded normal values in TBA hearts. Cardiac mass median Z-score in TBA hearts with pulmonary hypertension of III-IV grade was significantly higher if compared to pulmonary hypertension of I-II grade. Diameters of tricuspid and pulmonary valve were significantly bigger in TBA hearts without aortic obstruction.

Conclusions: There is an increase in cardiac mass, right ventricular myocardial thickness as well as pulmonary valve diameter in TBA hearts if compared to age adjusted normal values. The degree of pulmonary hypertension may contribute to the increase in cardiac mass.

Keywords: Taussig-bing; Morphometry; Cardiac mass; Cardiac valves

Introduction

Taussig-Bing is one of the variants of the double outlet right ventricle. This anomaly was initially described in 1949 as transposition of the great arteries [1]. In 1950, the similar case was reported, and the anomaly was eventually named “the Taussig-Bing heart” [2]. The characteristic feature of the Taussig-Bing anomaly (TBA) that makes it different from partial transposition of the great arteries (TGA) is sub pulmonary ventricular septal defect (VSD). This malformation was defined as “a double-outlet right ventricle with semilunar valves side-by-side and approximately at the same height, bilateral conus, and subpulmonary VSD” [3]. The term of “double-outlet right ventricle” (DORV) was initially used in 1952 [4] and introduced in its modern meaning in 1957 [5], that replaced the term of “partial transposition of the great arteries”. The most thoroughly used classification of DORV is based on anatomical position of VSD and the great arteries [6]. Per modern nomenclature, TBA is a DORV with subpulmonary VSD [7]. Morphological variability of TBA is guided by the relationship between the great arteries [8,9]. Moreover, TBA may be accompanied by intracardiac and extracardiac malformations that influence the parameters of the growing heart as well as natural history of the disease [10]. These malformations include aortic obstruction at different levels (narrowing of the left ventricular outflow tract, hypoplasia of the aortic arch, coarctation of the aorta), multiple VSDs, and others [11-13].

Morphometric studies of hearts with TBA are limited and in the majority of cases, do not contain quantitative assessment of intracardiac structures [3,14-16]. The aim of this study was to measure intracardiac structures (e.g. cardiac mass, diameter of valves, thickness of the myocardium), to assess their deviation from the individual norm, as well to determine how these parameters are influenced by anthropometric variability or pulmonary hypertension.

Material and Methods

The protocol of this retrospective study was approved by the Institutional Review Board of the Bakoulev Center for Cardiovascular surgery. Because of its retrospective nature, the study did not require a specific Informed Consent Form. However, all patients’ parents/guardians did sign the general Informed Consent Form that allowed using data received during their examination and surgical treatment for scientific analysis and publication as well using cadaver’s organs/tissues for research purposes.

Thirty pediatric hearts with TBA underwent morphometric assessment at the Bakoulev Center for Cardiovascular Surgery (Moscow, Russia) during the years 1998-2013. There were 20 male

*Corresponding author: Sergey B Zaets, 1 Wall Street, Apt. 5B, Fort Lee, NJ 07024, USA, Tel: 1-201-873-8901; E-mail: zaets001@yahoo.com

Received: August 22, 2014; Accepted: September 19, 2014; Published: September 21, 2014


Copyright: © 2014 Bockeria LA et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.
and 10 female hearts. The median age at the moment of death was 0.4 (95% confidence interval: 0.04; 9.1) years. All patients had pulmonary hypertension of grade I-II (n=24) or grade III-IV (n=6) per Heath-Edwards. Preparation of heart specimen was conducted using the methodology developed at the Bakoulev Center that allows preserving the original ratio of intracardiac structures [17]. Morphological criteria of TBA were as follows: 1). Both aorta (fully) and pulmonary artery (fully or predominantly) arise from the right ventricle; 2). Subpulmonary VSD is present; 3). Subaortic muscular conus is located in the right ventricle; 4). Subpulmonary conus or the left part of the bulboventricular fold (preserved or partially absorbed) is present (i.e. the mitral-semilunar fibrous continuation of the left ventricle is absent). Morphometric assessment included the measurement of the following parameters: cardiac mass diameter of the fibrous annulus of all cardiac valves (mitral, tricuspid, aortic, and pulmonary), myocardial thickness of the both right and left ventricle. Results of valvular morphometry were compared with individual normative data presented by Schulz DM and Giordano DA as mean values with standard deviation (SD) [18], and Z-score index was calculated.

Data are presented as median values and 95% confidence interval (CI) or mean and SD as appropriate. Differences between cardiac parameters in compared groups were determined by Student’s t test or ANOVA as appropriate. The Spearman rank correlation was used to reveal association between variables. The significance was set at the level of p<0.05. Statistical analysis was performed using SPSS computer software package (SPSS Inc., Chicago, IL).

Results

All hearts represented situs solitus with concordant atrioventricular junction. Morphologically right ventricle was located to the right from morphologically left ventricle. There was a levocardia. Both subpulmonary and subaortic conus were present. The great arteries originated from the right ventricle. Variants of the relationship between the major arteries were as follows: aortic valve located to the right and in front of the pulmonary valve (n=12); aortic valve located to the right and side-by-side from the pulmonary valve (n=5); aortic valve located to the right and behind the pulmonary valve (i.e. normal position of the great arteries) (n=13). VSD was always subpulmonary. Multiple VSDs were present in 8 cases. Aortic obstruction was revealed in 17 cases (Table 1). The anatomy of TBA heart is shown at the Figure 1.

The following quantitative signs that are typical for TBA were

<table>
<thead>
<tr>
<th>Single level obstruction (n=9)</th>
<th>Multi-level obstruction (n=8)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Coarctation of aorta – 3</td>
<td>Subaortic stenosis + interrupted aortic arch – 2</td>
</tr>
<tr>
<td>Subaortic stenosis – 1</td>
<td>Valvular aortic stenosis + coarctation of aorta – 2</td>
</tr>
<tr>
<td>Aortic arch hypoplasia – 2</td>
<td>Valvular aortic stenosis + subaortic stenosis + interrupted aortic arch – 1</td>
</tr>
<tr>
<td>Coarctation of aorta with aortic arch hypoplasia – 2</td>
<td>Valvular aortic stenosis + subaortic stenosis + aortic arch hypoplasia – 1</td>
</tr>
<tr>
<td>Valvular aortic stenosis – 1</td>
<td>Valvular aortic stenosis + aortic arch hypoplasia + coarctation of aorta – 1</td>
</tr>
</tbody>
</table>

*Stenosis was caused by the aneurism of non-coronary aortic leaflet

Table 1: Types of aortic obstructions in hearts with Taussig-Bing anomaly.
Cardiac mass in TBA hearts exceeded normal values in 87% of cases. In these hearts, the mean difference between their cardiac mass and individual normative values and Z-score reached 31.5g (CI: +12.1; 124.3) and +4.3 (CI: +2.3; +10.9), respectively. This difference increased with age. Cardiac mass indexed to the height or to the body weight also significantly exceeded normal values (Figure 2). Cardiac mass was increased predominantly because of the increase in the right ventricular mass. Thickness of the right ventricular myocardium significantly exceeded normal values, in contrast to normal thickness of the left ventricular myocardium (Table 2). Both cardiac mass and the difference between the cardiac mass and individual normative value correlated with the body surface area (r= 0.94; p<0.001, and r=0.76, p=0.004, respectively. Cardiac mass median Z-score in hearts with pulmonary hypertension of III-IV grade was significantly higher if compared with normal and TBA hearts (Table 4). Cardiac mass indexed to the body height *Normal values are given per Schulz DM, Giordano DA, 1962 [18].

Cardiac mass indexed to the body weight **Normal values are given per Falc AA, 1901 [23].

<table>
<thead>
<tr>
<th>Evaluated parameters</th>
<th>Morphometric values (mean ± SD or median with 95%CI)</th>
<th>Z-score (median with 95%CI)</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thickness of right ventricular myocardium (mm)</td>
<td>6.2 ± 2.4 (CI: 4.45; +18.3)</td>
<td>+2.6 ± 0.2 (CI: +0.5; +18.3)</td>
<td>0.001</td>
</tr>
<tr>
<td>Thickness of left ventricular myocardium (mm)</td>
<td>7.6 ± 2.4 (CI: +1.8; +2.9)</td>
<td>7.0 ± 1.4 (CI: +0.35; +1.8)</td>
<td>NS</td>
</tr>
<tr>
<td>Diameter of tricuspid valve (mm)</td>
<td>16.1 ± 4.4 (CI: +3.8; +2.6)</td>
<td>16.6 ± 4.4 (CI: +0.03; +2.6)</td>
<td>NS</td>
</tr>
<tr>
<td>Diameter of mitral valve (mm)</td>
<td>15.9 ± 4.2 (CI: +1.5; +4.5)</td>
<td>13.7 ± 4.1 (CI: +1.6; +2.0)</td>
<td>0.05</td>
</tr>
<tr>
<td>Diameter of pulmonary valve</td>
<td>13.3 ± 4.8 (CI: +1.8; +4.5)</td>
<td>9.8 ± 2.8 (CI: +0.60; +2.0)</td>
<td>NS</td>
</tr>
<tr>
<td>Diameter of aortic valve</td>
<td>10.4 ± 4.1 (CI: +1.8; +4.5)</td>
<td>9.4 ± 2.8 (CI: +0.60; +2.0)</td>
<td>NS</td>
</tr>
<tr>
<td>Cardiac mass (g)</td>
<td>66 (27.5; 230.8) (CI: +3.8; +10.4)</td>
<td>35.5 (21.0; 30.35) (CI: +3.8; +10.4)</td>
<td>0.004</td>
</tr>
</tbody>
</table>

*Normal values are given per Schultz DM and Giordano DA [18]. NS – non-significant

Table 2: Intracardiac parameters in hearts with Taussig-Bing anomaly (comparison with normal hearts).

Discussion

Only few publications report morphometric evaluation of hearts with TBA, which includes descriptive and quantitative assessment of cardiac preparations with four morphological types of TBA [16]. The authors have measured cardiac mass, wall thickness and volumes of both ventricles as well as circumference of atrioventricular and semilunar valves. Unfortunately, they have not provided actual numbers but only postulated the increase in cardiac mass and changes in dimensions of cardiac valves. Different parts of bilateral conus (e.g., width of subpulmonary conus between the pulmonary and mitral valve as well as between the aortic and tricuspid valve, length of “crista supraventricularis” that separates septal leaflets of both semilunar valves from the tricuspid valve) have been also measured [3]. However, this work does not provide any statistical analysis. Later on, the length of the conus septum as well as the distance between semilunar and atrioventricular valves has been assessed, and the relationship between the great arteries has been determined [14]. The main goal of these studies was to reveal anatomical features of ventricular outflow tracts in different forms of transposition of the great arteries and TBA. Many investigators believe that the position of the conus septum in TBA determines the type of VSD, significantly influences the hemodynamics, and eventually impacts the tactics of surgical treatment [15,19-22]. They postulate that the anterior shift

Figure 2A: Cardiac mass indexed to the body height *Normal values are given per Schulz DM, Giordano DA, 1962 [18].

B: Cardiac mass indexed to the body weight **Normal values are given per Falc AA, 1901 [23].
Further interpretation and mathematical analysis of received data will help to determine the predicted cardiac mass in TBA that may help in optimizing the dose of cardiac protective and inotropic medications in patients with this anomaly. The findings of the progressive increase in cardiac mass supports the approach of early surgical repair of TBA.

References