Anatomy & Physiology

Bockeria et al., Anat Physiol 2014, 4:4
http://dx.doi.org/10.4172/2161-0940.1000158

Research Article Open Access

Quantitative Anatomy of Taussig-Bing Anomaly

Leo A Bockeria[1], Osman A Makhachev[1,2], Margarita V Gorgeeva[1], Marina S Panova[1], Tatiana Yu Philippkina[1], Titalav Kh Khiriev[1] and Sergey B Zaets[1]

[1]Bakoulev Center for Cardiovascular Surgery, Moscow, Russia
[2]Dagestan 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 median 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 valvular 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 intra- 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. 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

Citation: Bockeria LA, Makhachev OA, Gorgeeva MV, Panova MS, Philippkina TYu, et al. (2014) Quantitative Anatomy of Taussig-Bing Anomaly. Anat Physiol 4: 158. doi:10.4172/2161-0940.1000158

Citation: Bockeria LA, Makhachev OA, Gorgeeva MV, Panova MS, Philippkina TYu, et al. (2014) Quantitative Anatomy of Taussig-Bing Anomaly. Anat Physiol 4: 158. doi:10.4172/2161-0940.1000158

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 bulboventriculr 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

Single level obstruction (n=9)	Multi-level obstruction (n=8)	
	Two-level obstruction (n=3)	Three-level obstruction (n=5)
Coarctation of aorta – 3 Subaortic stenosis – 1 Aortic arch hypoplasia – 2 Coarctation of aorta with aortic arch hypoplasia – 2 Valvular aortic stenosis – 1*	Subaortic stenosis + interrupted aortic arch – 2 Valvular aortic stenosis + aortic arch hypoplasia - 1	Valvular aortic stenosis + subaortic stenosis + coarctation of aorta – 2 Valvular aortic stenosis + subaortic stenosis + interrupted aortic arch – 1 Valvular aortic stenosis + subaortic stenosis + aortic arch hypoplasia – 1 Valvular aortic stenosis + aortic arch hypoplasia + coarctation of aorta - 1
*Stenosis was caused by the aneurism of non-coronary aortic leaflet		

Table 1: Types of aortic obstructions in hearts with Taussig-Bing anomaly.

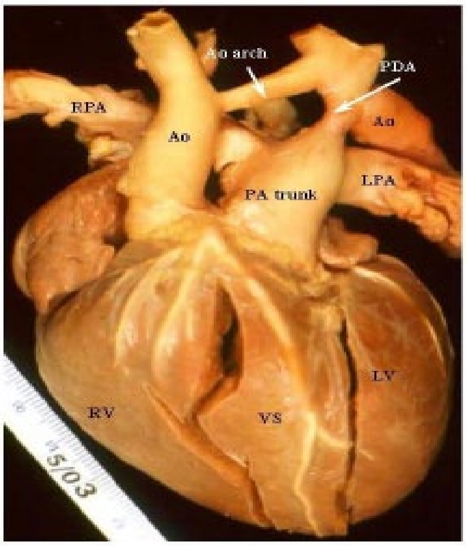

Figure 1a: Preparation of the heart with Taussig-Bing anomaly. General view of the heart. Segmental formula: [(S)C(DRH,L,SPC-SAC) DORV(D-P)]. Ao-aorta; LV – left ventricle; LPA – left pulmonary artery; PA – pulmonary artery; PDA – patent ductus arteriosus; RPA – right pulmonary artery; RV – right ventricle; VS – ventricular septum.

Figure 1b: Preparation of the heart with Taussig-Bing anomaly. Right ventricle, CS – conus septum; TV – tricuspid valve; VSD – ventricular septal defect.

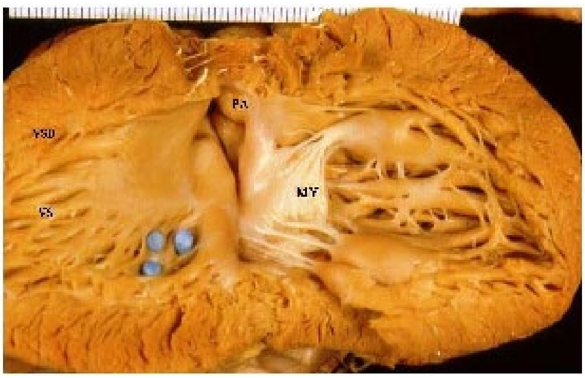

Figure 1c: Preparation of the heart with Taussig-Bing anomaly. Left ventricle, MV – mitral valve.

identified: cardiac mass, thickness of the right ventricular myocardium as well as the diameter of the pulmonary valve significantly exceeded normal values (Table 2).

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-IY grade was significantly higher if compared with pulmonary hypertension of I-II grade: +7.3 (CI: +0.4; +11.9) vs. +3.8 (CI: +0.26; +8.9), p<0.001. Gender, presence of aortic obstruction or multiple VSDs or position of the great arteries did not have any impact on cardiac mass (Table 3).

Evaluated parameters	Morphometric values (mean ± SD or median with 95%CI)		Z-score (median with 95%CI)	*p* value
	TBA	*Normal hearts**		
Thickness of right ventricular myocardium (mm)	6.2 ± 2.4	2.6 ± 0.2	+4.45 (+0.5; +18.3)	**0.001**
Thickness of left ventricular myocardium (mm)	7.6 ± 2.4	7.0 ± 1.4	+0.35 (-1.8; +2.9)	NS
Diameter of tricuspid valve (mm)	16.1 ± 4.4	16.6 ± 4.4	+0.03 (-3.8; +2.6)	NS
Diameter of mitral valve (mm)	15.9 ± 4.2	13.7 ± 4.1	+1.40 (-1.5; +4.5)	0.05
Diameter of pulmonary valve	13.3 ± 4.8	9.9 ± 2.8	+2.00 (-1.6; +5.8)	**0.002**
Diameter of aortic valve	10.4 ± 4.1	9.4 ± 2.8	+0.60 (-1.8; +4.5)	NS
Cardiac mass (g)	66 (27.5; 230.8)	35.5 (21.0; 30.35)	+3.80 (+0.2; +10.4)	**0.004**

*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).

In the majority of TBA hearts, diameter of the tricuspid as well as the aortic valve was within normal limits (in 81% and 68% of cases, respectively) (Table 2). The difference in mitral valve diameter between normal and TBA hearts was on the borderline of statistical significance. The mean diameter of the pulmonary valve in TBA group significantly exceeded normal values. Diameters of cardiac valves correlated with body surface area (tricuspid valve: r=0.83, p<0.001; mitral valve: r=0.87, p<0.001; aortic valve: r=0.98, p<0.001; pulmonary valve: r=0.85, p<0.001). Diameters of all cardiac valves were significantly bigger in TBA hearts with higher degree of pulmonary hypertension (Table 4). Diameters of tricuspid and pulmonary valve were significantly bigger in TBA hearts without aortic obstruction, whereas mitral and aortic valve did not significantly differ in diameter (Table 5).

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 statistial 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

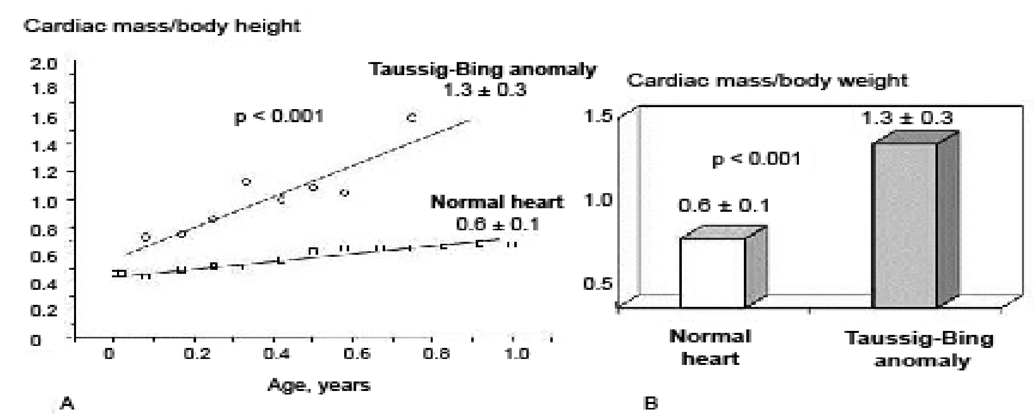

Figure 2A: Cardiac mass indexed to the body height *Normal values are given per Schulz DM, Giordano DA, 1962 [18].
B: Cardiac mass indexed to the body weight **Normal values are given per Falc AA, 1901 [23].

Parameters	Cardiac mass (g) median (95% CI)	*p* value
Gender Male gender (n=19) Female gender (n=11)	70 (22; 250) 55 (32; 200)	NS
Aortic obstruction Aortic obstruction (n=17) No aortic obstruction (n=13)	60 (22; 200) 73.5 (35; 250)	NS
Number of VSDs Single VSD (n=19) Multiple VSD (n=11)	65 (32; 250) 67 (22; 200)	NS
Position of great arteries (aortic and pulmonary valve) Aortic valve is anterior and to the right (n=15) Aortic valve is side-by-side and to the right (n=7) Aortic valve is posterior and to the right (n=8)	55 (22; 155) 110 (35; 200) 67 (32; 250)	NS
Severity of pulmonary hypertension Pulmonary hypertension grade I-II Pulmonary hypertension grade III-IV	55 (24; 237) 157.5 (100; 250)	**0.001**
NS – non-significant		

Table 3: Effect of different parameters on cardiac mass.

Cardiac valve	Pulmonary hypertension of I-II stage by Heath-Edwards (n=24)	Pulmonary hypertension of III-IY stage by Heath-Edwards (n=6)	*p* value
Tricuspid valve	14.7 ± 3.5	21.7 ± 6.1	0.001
Mitral valve	14.6 ± 3.3	20.7 ± 4.3	0.001
Pulmonary valve	12.0 ± 3.8	18.5 ± 5.0	0.001
Aortic valve	9.1 ± 2.8	15.2 ± 4.8	0.001

Table 4: Diameters of cardiac valves in patients with different stages of pulmonary hypertension.

Diameter of cardiac valves (mm)	Patients without aortic obstruction (n=13)	Patients with aortic obstruction (n=17)	*p* value
Tricuspid valve	18.6 ± 3.5	14.0 ± 3.2	0.04
Mitral valve	16.8 ± 4.7	15.7 ± 4.2	NS
Pulmonary valve	15.3 ± 5.6	11.9 ± 2.7	0.03
Aortic valve	12.2 ± 4.5	8.3 ± 1.7	NS
NS – non-significant			

Table 5: Diameters of cardiac valves in patients with and without aortic obstruction.

of the conus septum during embryogenesis leads to the development of subaortic stenosis, "overriding" pulmonary valve as well as to the formation of "malalignment" VSD. In turn, subaortic stenosis leads to the coarctation of aorta or to the interruption of the aortic arch.

Our study presents full quantitative assessment of TBA. Not only main cardiac structures and parameters such as diameters of valves, thickness of both ventricular walls, and cardiac mass have been measured, but also their comparison with corresponding normal values have been made and Z-score calculated. This comparison allowed assessing the degree of deviation from individual normal value. The study has revealed the 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 presence of subpulmonary VSD as well as biventricular position of the pulmonary valve in TBA contributes to the development of pulmonary hypertension that progress with age [22,23]. Our work has confirmed that the degree of pulmonary hypertension may contribute to the increase in cardiac mass, whereas gender, position of the great arteries, subaortic obstruction or multiple VSDs do not effect this parameter.

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 cardio 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

1. Taussig HB, Bing RJ (1949) Complete transposition of the aorta and a levoposition of the pulmonary artery; clinical, physiological, and pathological findings. Am Heart J 37: 551-559.
2. Lev M, Volk BW (1950) The Pathologic Anatomy Of The Taussig-Bing Heart: Riding Pulmonary Artery; Report Of A Case. Bull Int Assoc Med Mus 31: 54-64.
3. Van Praagh R (1968) What is the Taussig-Bing malformation? Circulation 38: 445-449.
4. Braun K, De Vries A, Feingold DS, Ehrenfeld NE, Feldman J, et al. (1952) Complete dextroposition of the aorta, pulmonary stenosis, interventricular septal defect, and patent foramen ovale. Am Heart J 43: 773-780.
5. Witham AC (1957) Double outlet right ventricle; a partial transposition complex. Am Heart J 53: 928-939.
6. Lev M, Bharati S, Meng CC, Liberthson RR, Paul MH, et al. (1972) A concept of double-outlet right ventricle. J Thorac Cardiovasc Surg 64: 271-281.
7. Walters HL 3rd1, Mavroudis C, Tchervenkov CI, Jacobs JP, Lacour-Gayet F, et al. (2000) Congenital Heart Surgery Nomenclature and Database Project: double outlet right ventricle. Ann Thorac Surg 69: S249-263.
8. Mavroudis C, Backer CL, Muster AJ, Rocchini AP, Rees AH, et al. (1996) Taussig-Bing anomaly: arterial switch versus Kawashima intraventricular repair. Ann Thorac Surg 61: 1330-1338.
9. Wilcox BR, Cook AC, Anderson RH (2006) Surgical Anatomy of the Heart. (3rd edn), Cambridge University Press, UK.
10. Alsoufi B, Cai S, Williams WG, Coles JG, Caldarone CA, et al. (2008) Improved results with single-stage total correction of Taussig-Bing anomaly. Eur J Cardiothorac Surg 33: 244-250.
11. Griselli M, McGuirk SP, Ko CS, Clarke AJ, Barron DJ, et al. (2007) Arterial switch operation in patients with Taussig-Bing anomaly--influence of staged repair and coronary anatomy on outcome. Eur J Cardiothorac Surg 31: 229-235.
12. Sinzobahamvya N, Blaschczok HC, Asfour B, Arenz C, Jussli MJ, et al. (2007) Right ventricular outflow tract obstruction after arterial switch operation for the Taussig-Bing heart. Eur J Cardiothorac Surg 31: 873-878.
13. Rodefeld MD, Ruzmetov M, Vijay P, Fiore AC, Turrentine MW, et al. (2007) Surgical results of arterial switch operation for Taussig-Bing anomaly: is position of the great arteries a risk factor? Ann Thorac Surg 83: 1451-1457.
14. Kurosawa H, Van Mierop LH (1986) Surgical anatomy of the infundibular septum in transposition of the great arteries with ventricular septal defect. J Thorac Cardiovasc Surg 91: 123-132.
15. Stellin G, Zuberbuhler JR, Anderson RH, Siewers RD (1987) The surgical anatomy of the Taussig-Bing malformation. J Thorac Cardiovasc Surg 93: 560-569.
16. Lev M, Rimoldi HJ, Eckner FA, Melhuish BP, Meng L, et al. (1966) The Taussig-Bing heart. Qualitative and quantitative anatomy. Arch Pathol 81: 24-35.
17. Sinev AF (2001) Basics of clinical anatomy and embryogenesis of the human heart. In: Lectures on cardiology. Bockeria LA, Ed: 171-185. Bakoulev Center for Cardiovascular Surgery, Moscow, Russia.
18. Schulz DM, Giordano DA (1962) Hearts of infants and children. Weights and measurements. Arch Pathol 74: 464-471.
19. Ueda M, Becker AE (1985) Classification of hearts with overriding aortic and pulmonary valves. Int J Cardiol 9: 357-369.
20. Anderson RH, Lenox CC, Zuberbuhler JR (1983) Morphology of ventricular septal defect associated with coarctation of aorta. Br Heart J 50: 176-181.
21. Waldman JD, Schneeweiss A, Edwards WD, Lamberti JJ, Shem-Tov A, et al. (1984) The obstructive subaortic conus. Circulation 70: 339-344.
22. Tuma S, Poshivalova V, Shovranyak Ya (1990) Infundibular septum in double outlet reght ventricle. In: Morphology and morphometry of the normal heart and hearts with congenital heart defects, Moscow, Russia.
23. Falc AA (1901) Cardiac growth in children of different age. Sankt Petersburg, Russia.