

A CASE OF RARE CONGENITAL HEART MALFORMATION: TAUSSIG- BING SYNDROME

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Abstract

Introduction- Taussig- Bing Syndrome is a rare congenital heart malformation that was first described by Helen Taussig and Richard Bing. The aim of this article is to show the main morphological changes in this case of rare congenital malformation. Performed methods were autopsy examination, clinical data analysis, x-ray examination and microscopic examination. Results- the index case is 34 days- old male newborn, born in 37 gestational age, with weight 2.150gr. and height- 45 cm. The heart was found enlarged with normal position in the chest. During dissection of the heart there were detected: hypertrophy and dilation of the right ventricle; the aorta arise entirely above the right ventricle; the dilated pulmonary valve does not override the left ventricular cavity; pulmonary artery arises primarily from the right ventricle and partially overrides the ventricular septum and a high and large sub- pulmonary ventricular septal defect. Conclusions- The treatment of this syndrome is continuing to be an actual medical problem. The right diagnose and the adequate treatment can widely reduce the mortality and morbidity from this disease.

Keywords: *Taussig- Bing syndrome, congenital heart malformation, autopsy*

Introduction:

The double outlet right ventricle (DORV) is a rare malformation that represents 1-5% of all congenital heart diseases. The Taussig-Bing syndrome is the second most frequent cause of DORV, just after the Fallot type, and it is characterized by transposition of the aorta to the right ventricle and malposition of the pulmonary artery with ventricular septal defect (VSD) under the pulmonary artery. This anomaly was initially described in 1949 by Taussig and Bing as transposition of the great arteries. The important difference between Taussig-Bing's anomaly and transposition of the great arteries were described by Richard Van Praagh in his editorial review and stressed that the absence of pulmonary-mitral continuity is the characteristic feature of this anomaly. Taussig and Bing emphasized that the overriding of pulmonary artery was an integral part of this malformation. This anomaly was eventually named as "Taussig-Bing heart" in 1950 by Lev and associates and subsequently introduced the spectrum of Taussig- Bing hearts depending on the degree of pulmonary artery overriding (right - sided, intermediate, and left sided). The deviation of infundibular septum as a fundamental requisite to define these hearts, collectively termed as "Taussig-Bing complex". This syndrome is a rare disease but still remains a challenging diagnosis, due to its occurrence at neonatal age of presentation and frequently nonspecific clinical symptoms. Despite improved diagnostic and surgical techniques, there are still a significant number of cases discovered only at the time of autopsy.

Case presentation:

We present a case of multi- malformative syndrome, which includes double- outlet right ventricle (DORV) associated with esophageal and anal atresia, hypospadias and testicular agenesis on the left side and renal hypoplasia on the right side, detected by postmortem examination of the corpse. Clinical documents with x-rays images, pathology reports and hematoxylin and eosin-stained samples were reviewed. The present case is a 34 days- old male newborn, from third normal pregnancy, born in 37 gestational age, with weight 2.150gr. and height- 45 cm., who had general cyanosis and petechial hemorrhages of the skin since the moment of birth. His echocardiography and electrocardiography showed an atrial flutter (220-230/min) (Fig.1 and Fig.2), with signs of right ventricular hypertrophy and dilation, pulmonary hypertension and isolated ventricular septal defect (VSD).

The enlarged heart was also seen in X-ray examination (Fig.3).

The medical history of the parents and the relatives was unavailable. Emergency surgery operations /colostomy and gastrostomy/ were performed, because of the esophageal and anal atresia and also many reoperations because of the stomach perforation. Therefore the patient was transferred to the Intensive Care Unit. For 20 days of intensive care he developed symptoms of bradycardia (80/min), episodes of apnoea and fever, then the patient suddenly died.

Autopsy findings - The heart was found enlarged with normal position into the thoracic cavity. During dissection of the heart there were detected: hypertrophy and dilation of the right ventricle; the aorta arised entirely above the right ventricle (Fig. 4A); the dilated pulmonary valve did not override the left ventricular cavity; pulmonary artery arised primarily from the right ventricle and partially overrode the ventricular septum (Fig. 5C/D) and a large and high located sub- pulmonary ventricular septal defect was also presented (Fig. 4B). However, the ventricular septal defect was not confluent with the pulmonary valve, the defect was somewhat separated from the pulmonary valve by the sub- pulmonary and sub- aortic conal free wall musculature, forming the roof of the VSD.

The microscopic examination of the myocardium showed normal histological features (Fig.6)

Discussion:

Taussig-Bing malformation consists of transposition of the aorta to the right ventricle and malposition of the pulmonary artery with sub- pulmonary ventricular septal defect (1, 3). The original description of the Taussig-Bing anomaly has been broadened to include all kinds of double-outlet right ventricles with sub- pulmonary VSD (1, 2, 4). Anatomic repair of the Taussig-Bing anomaly is achieved with connection of the morphologically “left” ventricle to the aorta and of the morphologically “right” ventricle to the pulmonary artery (1, 2, 5, 6, 7). In selected patients, such anatomic correction can be achieved by inter- ventricular repair, also known as the Kawashima operation (1, 8, 9). The arterial switch operation, however, appears to be preferred. Excellent early and intermediate survival has been reported recently. Historically, the arterial switch operation has been associated with higher rates of morbidity and mortality (3) when it is performed in patients with the Taussig-Bing anomaly, compared with patients who undergo arterial switch for d-transposition of the great arteries. In children with Taussig-Bing anomaly, sub-aortic right ventricular outflow tract obstruction is found in 50% to 60% of the patients, and arch

obstruction is common and present in 39% to 52% of the cases. One of the earliest studies, which show that a Taussig-Bing anomaly, with or without arch obstruction, can be repaired via the arterial switch operation during the neonatal period with good outcome is reported in 1996 by investigators at the Royal Children's Hospital in Melbourne, Australia. This report describes 28 consecutive patients who underwent arterial switch operations with baffling of the left ventricle to the neo-aorta between 1983 and 1995. The overall operative mortality rate was 7% (2 of 28 patients). In 11 children with Taussig-Bing anomaly associated with aortic arch obstruction, the actuarial survival rate was 100%, and the freedom-from-reoperation rate was 73% at 6 years. The corresponding rates for 17 children with isolated Taussig-Bing anomaly were 81% and 100%. A recent report from the Hospital for Sick Children in Toronto describes 33 consecutive neonates with Taussig-Bing anomaly who underwent arterial switch operations between 1979 and 2005 (1, 10). The mean 1-year survival rate of 17 patients who were operated upon from 1979 through 1999 was only 47% \pm 5%, but it was 100% for 16 who were operated upon from 1999 through 2005. The 5-year event-free survival rates for those groups were 35% \pm 6% and 87% \pm 1%, respectively (1, 10).

Conclusion:

Taussig- Bing syndrome is a rare congenital malformation, treatment of which is continuing to be an actual medical problem. The correction of this congenital heart disease still represents a huge challenge for the health professionals. The timely diagnose and the adequate treatment can widely reduce the mortality and morbidity from this disease, and also can give better quality of life to all those children who are in a waiting list to cardiac surgery. This case report could help many medical workers from different specialties correctly to recognize this heart anomaly, which also could reduce the mortality and morbidity from this congenital disease.

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LEGEND:

Figure 1 and Figure 2. ECG, showing atrial flutter.

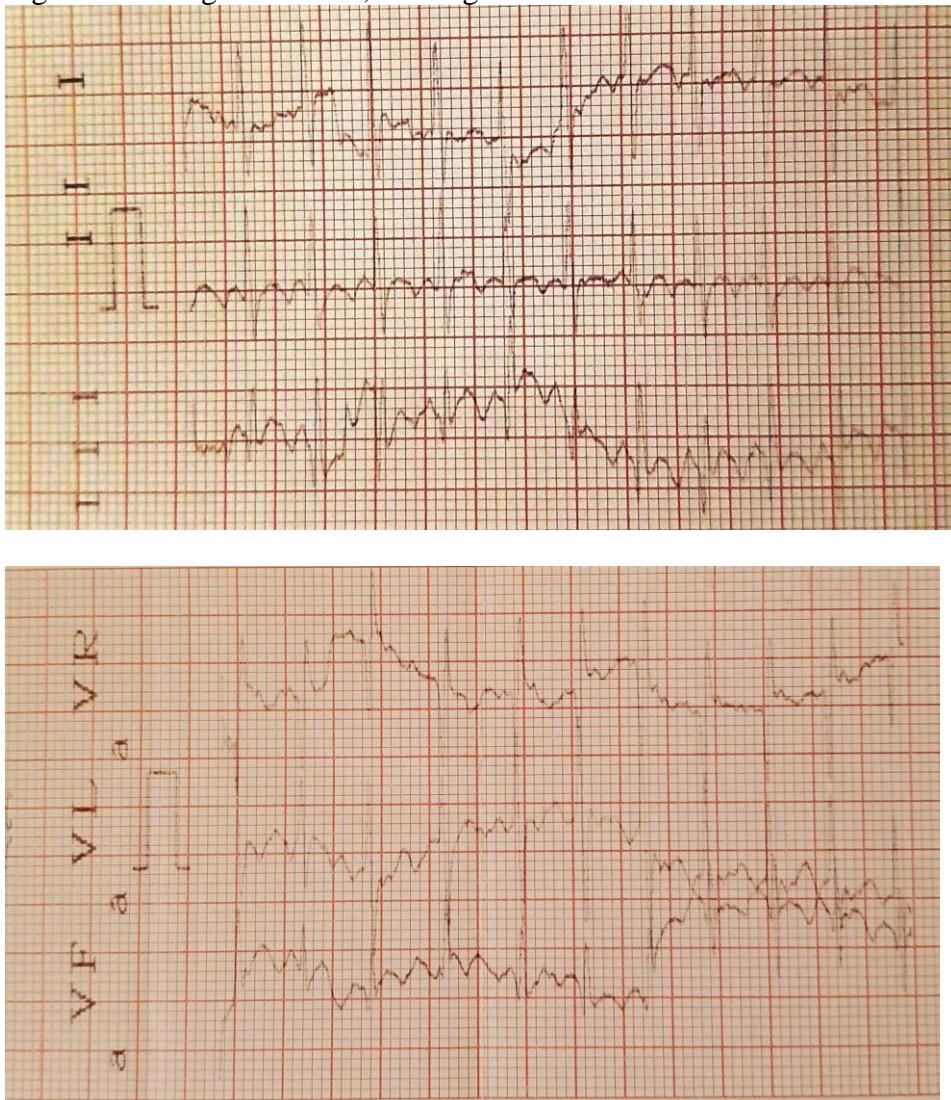


Figure 3. X-ray examination of the chest, showing enlargement of the heart.

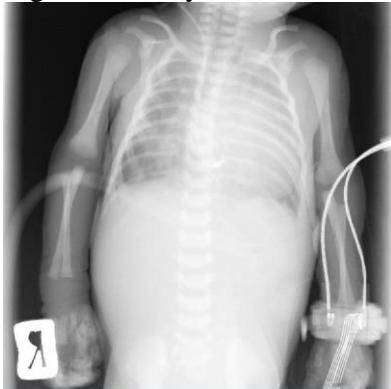
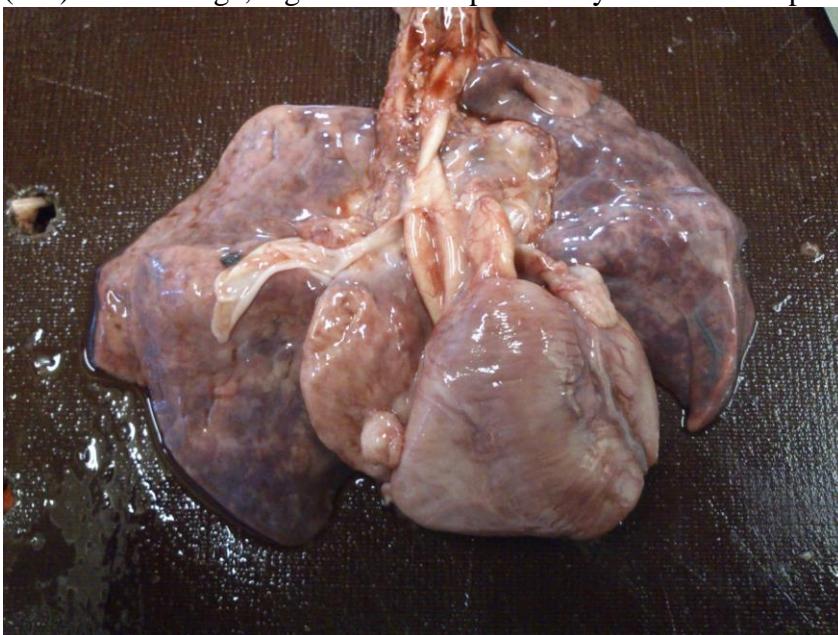


Figure 4. A) On the left are presented the lungs with right atrium (RA) and right ventricle (RV). B) On the right are presented the open right ventricle (RV), opened inner surface of the aorta (AA) and the large, high located subpulmonary ventricular septal defect (VSD).



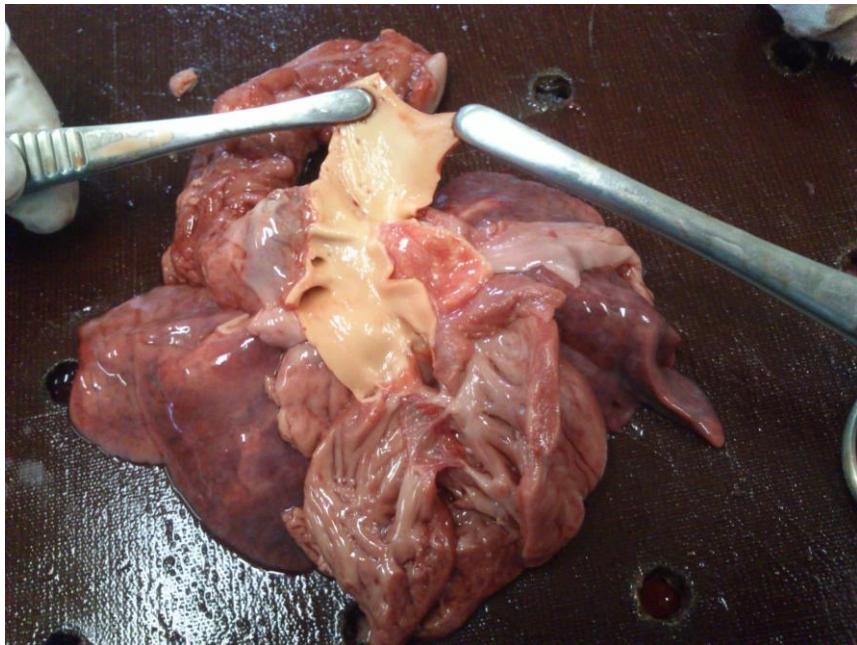
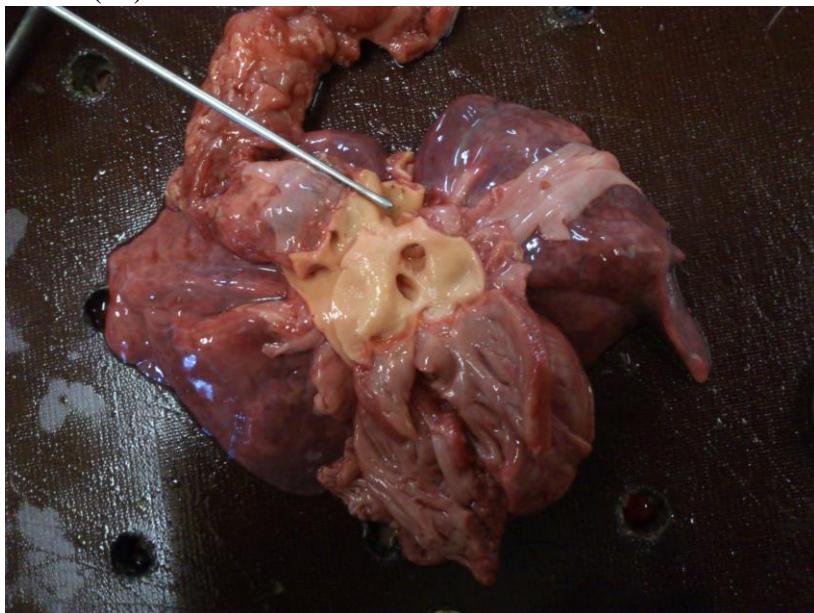


Figure 5. C) and D) On the left are presented the open pulmonary artery, which arises from right ventricule. On the right photo we can see the opened pulmonary artery and ventricular septal defect (*).



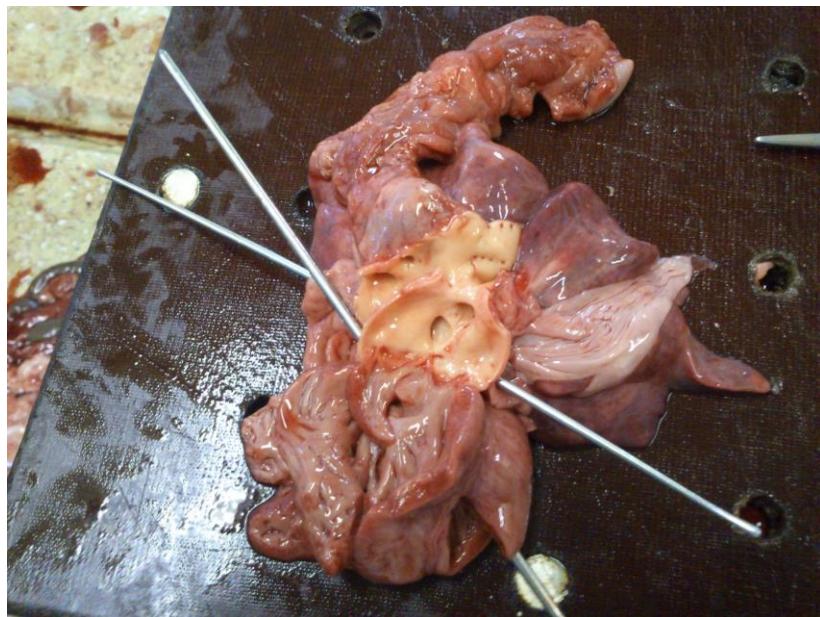


Figure 6. The myocardium showed normal histological features. H&E x40.

