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## Original scientific paper



## Profesional paper



## Review



## Case report

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# *Betimi i Hipokratit*

*Në çastin kur po hy në radhët e anëtarëve të profesionit mjekësor premtoj solemnisht se jetën time do ta vë në shërbim të humanitetit. Ndaj mësuesve do ta ruaj mirënjohjen dhe respektin e duhur. Profesionin tim do ta ushtroj me ndërgjegje e me dinjitet. Shëndeti i pacientit tim do të jetë brenga ime më e madhe. Do t'i respektoj e do t'i ruaj fshehtësitë e atij që do të më rrëfehet. Do ta ruaj me të gjitha forcat e mia nderin e traditës fisnike të profesionit të mjekësisë.*

*Kolegët e mi do t'i konsideroj si vëllezër të mi.*

*Në ushtrimin e profesionit ndaj të sëmurit tek unë nuk do të ndikojë përkatësia e besimit, e nacionalitetit, e racës, e politikës, apo përkatësia klasore. Që nga fillimi do ta ruaj jetën e njeriut në mënyrë apsolute. As në kushtet e kërcënimit nuk do të lejoj të keqpërdoren njohuritë e mia mjekësore që do të ishin në kundërshtim me ligjet e humanitetit. Këtë premtim po e jap në mënyrë solemne e të lirë, duke u mbështetur në nderin tim personal.*

# *The Oath of Hippocrates*

*Upon having conferred on me the high calling of physician and entering medical practice, I do solemnly pledge myself to consecrate my life to the service of humanity. I will give my teachers the respect and gratitude which is their due. I will practice my profession with conscience and dignity. The health of my patient will be my first consideration. I will respect the secrets which are confided in me, even after the patient has died. I will maintain by all the means in my power, the honor and the noble traditions of the medical profession.*

*My colleagues will be my brothers.*

*I will not permit considerations of religion, nationality, race, party politics or social standing to intervene between my duty and my patient. I will maintain the utmost respect for human life from its beginning even under threat and I will not use my medical knowledge contrary to the laws of humanity. I make these promises solemnly, freely and upon my honor*

Editorial Board

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# PRENATAL DIAGNOSIS OF TAUSSIG BING ANOMALY WITH COARCTATION OF THE AORTA - CASE REPORT

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## ABSTRACT

Taussing Bing anomaly is a rare congenital heart malformation. Congenital heart defects (CHDs) represent a formidable global health challenge, frequently necessitating intricate surgical interventions, particularly in the vulnerable populations of neonates and infants. Among these, anomalies involving the great arteries and ventricular septal defects (VSDs) pose unique difficulties due to their complex anatomy and profound physiological impact on the developing cardiovascular system.

We present a case of a fetal cardiac anomaly initially diagnosed as transposition of the great arteries (TGA) with subsequent identification of coarctation of the thoracic aorta (CoA). Post-termination autopsy confirmed Taussig Bing anomaly (TBA), a rare conotruncal malformation characterized by a double-outlet right ventricle (DORV) with subpulmonary ventricular septal defect (VSD) and malposition of the great arteries. This case highlights the diagnostic challenges in prenatal imaging and the importance of multidisciplinary evaluation in complex congenital heart disease (CHD).

## INTRODUCTION

Taussig Bing anomaly is a rare congenital cardiac defect accounting for <1% of CHD cases. It involves a DORV with a subpulmonary VSD and side-by-side great arteries, often associated with aortic arch anomalies (e.g., coarctation or interruption)(1). Prenatal diagnosis is challenging due to similarities with TGA and other conotruncal defects. We discuss a case initially suspected as TGA accompanied with CoA, later confirmed as TBA.

Taussig Bing anomaly carries a high mortality rate if left untreated, with most infants developing severe cyanosis and heart failure shortly after birth. Without intervention the mortality in the first year of life rises up to >80% due to hypoxia, pulmonary hypertension, or circulatory collapse. Neonatal survival depends on the degree of pulmonary stenosis which protects against pulmonary overcirculation, aortic arch obstruction (coarctation) which worsens systemic perfusion and other associated defects (VSD size, coronary anomalies)(2).

The primary goal of the surgical management strategies is anatomic correction to establish normal circulation(3). Approaches depend on anatomy and clinical status of the fetus and the possibilities whether or not the anomaly is compatible with life(4).

The etiology of congenital heart defects can be multifactorial, genetical, environmental, viral but also, sometimes not detected, therefore is a field of interest for many decades(1).

The association with other subsequent abnormalities such as pulmonary valve stenosis or coarctation of the aorta contribute for greater challenges prior to diagnose and the evaluation of the further management needs multidisciplinary approach with not rarely unfavorable outcome. Observations on anatomical associations and early surgical outcomes laid foundational groundwork for subsequent advancements in the field, cementing its status as a valuable historical benchmark in pediatric cardiology and cardiothoracic surgery.

A complete medical team consisted of obstetritian - fetal ehocardiographist, pediatritian, cardiac children surgeons, cardiologists, radiologist should focus on early diagnosis during pregnancy evaluation of the degree of damage and the rate of postpartal survival and also the quality of life. If prognosis is poor and worsens in utero, the pregnancy according to the doctors team should be terminated. If the prognosis is satisfactory, employing the correct surgical technique and determining the optimal timing for each case of this heart anomaly improves the patients' outcome(2).

## PURPOSE

The purpose of this article is to emphasize the importance of prenatal diagnosis in patients who have fetuses with congenital anomalies and the significance of ultrasound screening for diagnosing complex cardiomyopahies. In this case the prenatal diagnosis of Taussig Bing anomaly with coarctation of the aorta is represented.

## MATERIAL AND METHOD

A 30-year-old patient with first pregnancy came at the University Clinic of Gynecology and Obstetrics in Skopje, Republic of North Macedonia. She had her regular examinations until 21 gestational weeks. The patient was given questionnaire for history of diseases, for previous pregnancies and for other comorbidities. She was examined in detail. No previous comorbidities nor operations were detected. Screening for fetal anomalies was performed in Sistina Hospital. Fetal heart was observed in details and the great heart vessels suspected parallel due to transposition of great arteries. Fetal cardiologist confirmed the diagnose. For determining the outcome of the pregnancy, the patient was sent in Sofia, Bulgaria in referent cardiodiagnostic pediatric center. With multidisciplinary approach, by obstetrician, pediatrician and cardiac surgeon, the fetus was evaluated and complex cardiopathy was concluded on ultrasound with parallel great vessels and discordance in their lumen, mild VSD in the membranous part and coarctation of the aorta in its thoracic part. Their suspicion was the Taussig Bing anomaly. After detailed research they came to a conclusion that the outcome of the fetus is incompatible for life. With prolonging the pregnancy with the coarctation of the aorta there persisted a risk of dilatational ventriculomegaly which cannot be treatable with the operative techniques. Also, if the child would have been born the quality of life was evaluated poor with operational plan on every three months with a risk of low survival rate. Therefore, they suggested termination of pregnancy. Amniotic fluid was collected and send for karyotype analysis. After signed consent for terminating the pregnancy, induction was performed. The fetus and the placenta were send to the Institute of Pathology and histology for autopsy. Material for genetic analyses was collected from the fetus and both of the parents and sent in MANU for confirming the gene corresponding to cardiomyopathy syndrome. Blood sample for TORCH analyses were also collected.

## RESULTS AND DISCUSSION

From the used materials and methods, important information were obtained from the situation of the patient. It had been discovered that there was no consanguinity among both partners. Genetic panel testing which included 150 hereditary cardiomyopathy resulted normal. In the pregnancy, the patient underwent an ultrasound examination at 21.4 weeks of gestation (screening in the second trimester). In the ultrasound examination complex cardiopathy was diagnosed with initial findings of suspected TGA due to parallel great vessels on fetal echocardiography (figure 1). Further evaluation revealed narrowing of the aortic arch (CoA).

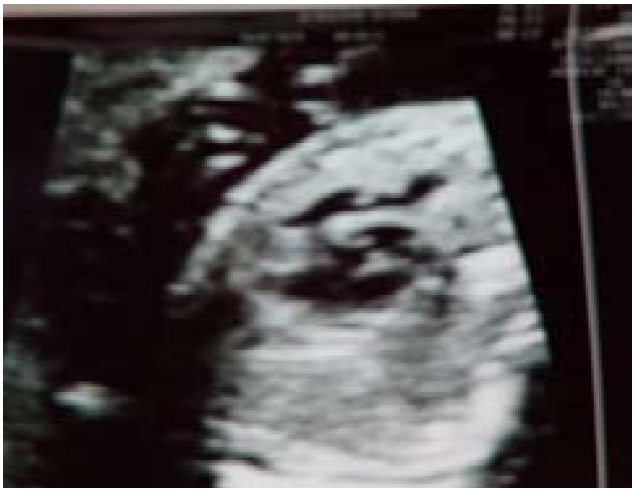

Figure 1. Ultrasound examination of complex cardiopathy

Because of these findings that indicate poor prognostic outcome, the pregnancy needed to be terminated. Natrium cloride-induced termination of the pregnancy was indicated for the pregnant patient, who gave birth to her malformed baby.

Figure 2. Examination of the fetus with Taussig – Bing anomaly

The autopsy findings confirmed Taussig Bing associated with double outlet right ventricle, subpulmonary VSD, malposed great arteries (aorta anterior and right of pulmonary artery 3 mm wide) and coarctation of the thoracic aorta (to 1 mm).

The results of the viral panel confirmed for rubella viral infection with high avidity confirmed in the first trimester. The severity of heart defects can vary depending on the timing, earlier infections generally lead to more severe outcome. Therefore, congenital cardiomyopathy due to acquired viral infection had the highest probability for this rare occurrence.

There persist diagnostic challenges between TGA vs. TBA. The diagnose of transposition of the great arteries is made according to the finding that aorta arises from the right ventricle (RV), pulmonary artery (PA) from the left ventricle (LV) whereas at Taussig Bing anomaly both great arteries arise from RV, but PA overrides the VSD. Misdiagnosis occurs due to similar vessel orientation. That is why the role of advanced imaging as fetal echocardiography may miss subtle anatomical differences. Postnatal and postmortem MRI/CT improves diagnostic accuracy. Associated anomalies like coarctation of the aorta (present in ~50% of TBA cases) worsens prognosis. Early detection is crucial for surgical planning(5.6).

According to literature review, TBA has a high mortality without surgical correction (neonatal palliation and eventual arterial switch or Rastelli procedure). Few procedures for neonatal stabilization (palliation before repair) include - usage of prostaglandin E1 (PGE1) which maintains ductal patency if systemic circulation depends on the PDA (critical in cases with coarctation). Also, balloon atrial septostomy (BAS) is recommended, if restrictive atrial septation worsens cyanosis, and pulmonary artery banding (PAB) bit it is rarely used, reserved for severe pulmonary overcirculation. Cyanosis is a consequence of poor mixing (due to restrictive atrial septum/VSD), dynamic LVOT obstruction, or pulmonary hypertension(4).

Definitive surgical repair that are proceeded are: arterial switch operation (ASO) and VSD closure is preferred when feasible (adequate LV pressure, favorable coronary anatomy). Survival at 5 years is 85–90%, but risks include coronary insufficiency and neo-aortic regurgitation. Rastelli procedure is chosen for significant LVOT obstruction; it involves baffling the VSD to the aorta and placing an RV-PA conduit. Ten-year survival is ~75%, but conduit replacements are often needed. Double Root Translocation (DRT) is an emerging alternative for complex coronary anatomy, with promising mid-term outcomes. Coarctation management of simultaneous repair during ASO (end-to-end anastomosis or patch augmentation) is preferred, but a staged approach (initial coarctation repair via thoracotomy, followed by ASO) may be used if needed. The survival rate is aproximatelly 80% at 10 years as long-term outcome but is probably associated with late complications that involve neo-aortic regurgitation (post-ASO), RVOT obstruction (post-Rastelli), and arrhythmias.

Reinterventions are necessary and are not rare, about 30–40% require reoperations (conduit replacement, residual VSD closure) or catheter-based interventions (stenting for recoarctation, PA dilation). Evaluation of the prognosis of the neoanate is crutial for continuing or terminating the pregnancy. Prenatal diagnosis improves the outcome (allowing planned delivery at a tertiary center) but accompanied poor prognostic factors such as coarctation of the aorta and small VSD (restricts systemic flow) and coronary anomalies (increases ASO complexity)(3,7).

A study by Konstantinov summarized surgical experiences, procedures, and patient outcomes. Four critically ill patients with Taussig Bing heart and coarctation underwent initial coarctation repair and pulmonary artery banding at a very young age (2 to 7 days of age). This highlights the acute need for palliation in these severely affected neonates. Two other patients with Taussig Bing did not require immediate intervention for coarctation as it was not hemodynamically significant, indicating a spectrum of severity. Five of the six patients with Taussig Bing and coarctation later underwent a Senning procedure, an atrial switch operation for transposition of the great arteries, between the ages of 7 weeks and 3.5 years. Surgeons recognized that aggressive, single-stage complete repairs, especially those involving significant myocardial incision (like right ventriculotomy) in fragile neonates, carried unacceptable risks. It highlights the importance of minimizing iatrogenic myocardial injury in the developing heart and adapting surgical strategy to the physiological resilience of the patient, a principle that continues to guide complex pediatric cardiac interventions today(8).

Parr et al. definitively showed a 53% coarctation rate in Taussig Bing versus 6% in TGA with VSD, a robust and statistically significant finding. Today, clinicians have access to high-resolution imaging modalities. While advanced imaging can confirm the diagnosis, the knowledge of this strong association from foundational papers like Parr et al. allows clinicians to form a high index of suspicion. If a complex transposition-like anomaly is suspected, the presence or absence of coarctation immediately narrows the differential diagnosis, even before definitive imaging results are fully interpreted. Continued vigilance for coarctation is needed in any neonate or infant presenting with a suspected transposition complex, particularly with features suggestive of double-outlet right ventricle or right ventricular outflow tract obstruction, a thorough and meticulous evaluation for coarctation of the aorta or other aortic arch anomalies is crucial. The high prevalence noted by Parr et al. makes this an essential diagnostic consideration that can significantly influence subsequent management(9,10).

While modern series often report excellent outcomes for one-stage repair, comparative studies on the long-term neurodevelopmental and cardiac functional outcomes of staged approaches (as initially advocated by Parr et al.) versus primary one-stage repairs in specific subsets of Taussig Bing patients (e.g., very low birth weight, extreme prematurity, or those with additional severe comorbidities) could provide valuable insights into optimal management strategies for the most challenging cases(11).

Continued research into how advanced imaging modalities (e.g., 3D printing of patient-specific cardiac models from imaging data, virtual reality surgical planning) can further mitigate the anatomical challenges highlighted by Parr et al. (e.g., difficult VSD closure) and improve surgical precision, potentially enabling safer definitive repairs in younger, more fragile infants.

Further research into the genetic and developmental pathways that lead to the strong association between Taussig-Bing malformation and coarctation could offer deeper insights into the pathogenesis of these complex CHDs. This could potentially lead to earlier diagnostic markers, non-invasive screening methods, or even novel preventative strategies based on developmental biology(11).

## CONCLUSION

This case underscores the complexity of diagnosing TBA prenatally. A high index of suspicion for conotruncal anomalies is essential when TGA is suspected, particularly with coexisting aortic arch obstruction. Multidisciplinary collaboration (fetal cardiology, perinatology, pathology) is critical for accurate diagnosis and counseling.

TBA requires early surgical correction, ideally with ASO when feasible. Coarctation complicates management, often requiring ductal stabilization and arch repair. Cyanosis is managed with BAS and PGE1, while long-term survival depends on monitoring for late complications. A multidisciplinary approach (cardiology, cardiac surgery, ICU) is essential for optimal outcomes.

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