# Clinical management of bloodless surgery for aortic valves fibroelastoma and coronary artery disease in a Jehova Witness

Yehova Şahidi'nde aort kapak fibroelastoması ve koroner arter hastalığının kansız ameliyatının klinik tedavisi

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#### ABSTRACT

A papillary fibroelastoma is an extremely rare cardiac tumor which often presents asymptomatically. However, it may lead to potentially severe complications due to thromboembolic risk. Cardiac surgery in Jehovah's Witnesses is a still infrequent procedure due to their religious beliefs which prohibit blood and blood product transfusion in any case, even in fatal emergency operations. Herein, we report a clinical management approach of blood preservation protocol during successful surgical treatment of coronary artery disease and a concomitant aortic valve fibroelastoma in a Jehovah's Witness in the light of literature data.

*Keywords:* Blood preservation; Jehovah's Witness; papillary fibroelastoma; transfusion.

Both during cardiac surgery and cardiopulmonary bypass (CPB) and in the postoperative period, a high rate of blood or blood product transfusion may be required. However, Jehovah's Witnesses (JWs) consider taking primary blood products (whole blood, packed red cells, white cells, platelets and plasma) as an act of violation against their religious beliefs in any case, even in fatal emergencies.<sup>[1,2]</sup>

A cardiac papillary fibroelastoma (CPF) is a benign endocardial papilloma, which not only the second most often primary cardiac tumor, but also

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Papiller fibroelastom, sıklıkla asemptomatik seyreden, çok nadir bir kalp tümörüdür. Ancak, tromboemboli riski nedeniyle muhtemel ciddi komplikasyonlara yol açabilir. Yehova Şahitlerinde kalp cerrahisi, ölümcül acil ameliyatlar dahil, her türlü durumda kan ve kan ürünü transfüzyonunu yasaklayan dini inançları gereği, halen seyrek bir işlemdir. Bu yazıda, bir Yehova Şahidi'nde koroner arter hastalığı ve eşlik eden aort kapak fibroelastomunun başarılı cerrahi tedavisi sırasındaki kan koruma protokolünün klinik tedavi yaklaşımı literatür verileri esliğinde sunuldu.

Anahtar sözcükler: Kan koruma; Yehova Şahidi; papiller fibroelastom; kan nakli.

the most prevalent valvular tumor.<sup>[3]</sup> The aortic valve is most often affected valve.<sup>[3]</sup> Although CPFs are frequently asymptomatic, they may cause ischemic or thromboembolic symptoms.<sup>[3]</sup> Herein, we present a bloodless approach for the successful surgical treatment of coronary artery disease with a rare cardiac tumor in a Jehovah's Witness.

## **CASE REPORT**

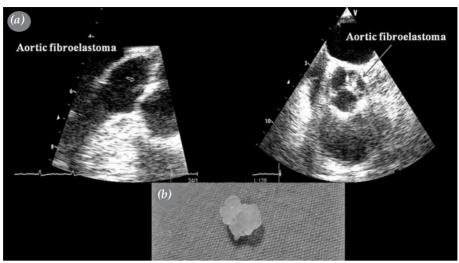
A 59-year-old male was referred to our hospital with a history of transient ischemic attack and a



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**Figure 1.** (a) A preoperative echocardiographic image of an intracardiac mass on the right coronary cusp of aortic valves. (b) Gross appearance of a multi-lobulated intracardiac mass (10x15x12 mm).

suspected mobile mass on the aortic valve. His medical history revealed that he experienced both transient left-sided hemiplegia (3-4/5) and diplopia two weeks ago. On physical examination, heart rhythm was in sinus with a 2/6 systolic murmur in the aortic area. Transthoracic echocardiography and transesophageal echocardiography revealed a mobile mass of 0.7 cm in diameter, which was attached to the right coronary cusp of the aortic valve, extending to the aorta (Figure 1). Other valves and left ventricle functioned properly. Inflammation markers such as C-reactive protein and sedimentation rate were normal. Electrocardiography, Chest X-ray, and laboratory tests produced normal findings. Hemoglobin level which was at the lower limit of normal was 120 g/L (male references, range: 129 to 159 g/L). Coronary angiography showed critical lesions of the left anterior descending (LAD) artery and circumflex artery (Figure 2). Due to the religious beliefs of the patient, the timing of surgery was planned according to the target value of hemoglobin level (140 g/L) and administration of ferrous sulfate, iron supplementation, and 600 U/kg of erythropoietin alpha (EPREX-Janssen-Cilag, Beerse, Belgium) subcutaneously every week for four weeks before surgery. The fourth dose was administered on the day of surgery. Preoperatively, hemoglobin level was 13.4 g/dL with a hematocrit value of 41.2 mL/dL. Autologous transfusion as 1 unit was prepared during the initial operation period.

The needed high care was provided to ensure minimal blood loss during surgery. The left internal mammary artery and great saphenous vein were prepared and, then, total CPB was established through the aorta and two-stage venous cannulation. The arterial and venous lines were shortened maximally and the amount of prime solution in reservoir was gently minimized with retrograde autologous priming. It consists of a pump in a closed circuit. The circuit is filled with the blood of the patient drained passively through a venous cannula, thereby, eliminating the crystalloid solutions, which refills the circuit and results in hemodilution. With this technique, hemoglobin level was 10.6 g/dL with a hematocrit value of 32.4 mL/dL during the initial circuit period. In addition, using the Cell Saver® system (Haemonetics Corporation, Braintree, MA, USA), an indirect re-transfusion system was applied for continuous autotransfusion for efficient

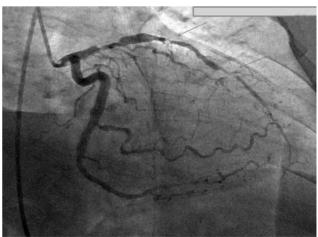


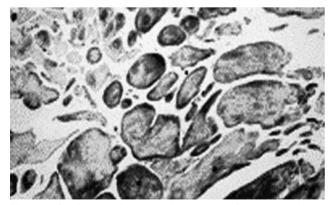
Figure 2. An angiography image of coronary lesions associated with an aortic fibroelastoma.

blood salvage in our patient who was willing to allow this procedure. First, aortic valve exploration was performed in a standard fashion using a valve sparing technique with a simple shave excision of the tumor. Then, the internal thoracic artery was sequentially anastomosed to the LAD as jumping and saphenous vein graft was anastomosed to the first obtuse marginal branch, as well. Controlled hypotension (the mean arterial pressure 55 to 60 mmHg) was also used to reduce intraoperative blood loss. Throughout CPB, hemoglobin level was preserved about 10 g/dL with a hematocrit value of 28 mL/dL. Cross clamping time was 52 minutes, while CPB time was 80 minutes. On termination of CPB, the residual volume of 350 mL from the reservoir and CPB connection lines was given via an aortic cannula. Postoperative course was uneventful. The amount of total drainage was 300 mL and the patient was transferred to the ward 18 hours after surgery. He was discharged on the postoperative seventh day with a hemoglobin level of 7.8 g/dL and a hematocrit value of 24.7 mL/dL.

Histopathological examination of the mass was reported as a papillary fibroelastoma containing papillary proliferation with a few fibroblasts and collagenous tissues covered with endothelial cells (Figure 3).

## **DISCUSSION**

The world JWs argue that the state has an interest in the preservation of life which is not absolute. <sup>[2]</sup> They claim that individuals have the right to control themselves and part of that autonomy is the right to make choices pertaining to one's health including the right to refuse unwanted medical treatment. <sup>[4]</sup> Although it should encompass all medical choices including to refuse



**Figure 3.** Histological specimens showing a papillary fibroelastoma containing papillary proliferation including a few fibroblasts and collagenous tissues covered with endothelial cells (H-E x 100).

blood products. According to the recent demographic statements in August 2012, there are nearly 2.5 thousands JWs in Turkey and about 7.53 million individuals have a membership to be actively engaged in preaching over 230 countries.[3] However, about 1,000 JWs die every year, as they refuse blood transfusion in the USA.<sup>[5]</sup> On the other hand, allogeneic blood transfusion is often necessary in cardiac surgery, although it increases mortality, morbidity, and major adverse outcomes.[6] Autologous blood and blood product transfusion should be a general approach for blood transfusions thanks to their relatively low risk. Next, the costs of red blood cell (RBC) transfusions have been underestimated, even when the cost of treatment of these adverse outcomes or prolonged stay in the intensive care unit and internalization time are excluded.[6,7] However, most surgeons are unaware of such problems. Blood management comprises three main elements: correction of perioperative anemia, minimizing perioperative blood-loss, and using low hemoglobin-based transfusion triggers. Indeed, blood transfusion for JWs is a medicolegal issue. During or after operation, even in the emergency setting, the physician who promises to the patient about he would never use blood transfusion into the body may experience the dilemma of the patient's consents or legal and professional requirements.

In some countries such as in France, where the patients refuse the use of blood transfusion, their consent is not taken under consideration during emergency, despite of a signed form of denial of blood transfusion. This implies that French rules are against the patient's consents. In such cases, medical ethics takes its course. On the other hand, in the USA, blood preservation protocol for JWs undergoing cardiac surgery have shown successful results.<sup>[4]</sup>

In accordance with the Turkish Penal Code,<sup>[8]</sup> exercise of the right and consent of the concerned is organized in the Article 26/1-2 in which no one can be penalized for exercising his right (1) and in case of consent regarding a right of which a person has full disposal, no one can be penalized (2).

This is why the multidisciplinary clinical team involves a cardiac surgeon, an anesthetist, and a hematologist. If it is agreed that surgery holds the best outcome for the patient, the JW patient must sign a form which explicitly expresses to refuse the treatment of primary blood components, called the Advanced Decision to Refuse Specified Medical Treatment, if he/she is willing to undergo the bloodless procedure. Once the patient signs the document, administration of blood products is deemed as unlawful, representing

an assault or the tort of trespass to the person and the surgeon is, then, liable to the criminal or civil proceedings.<sup>[9]</sup> Second, the opinion of the head surgeon should be obtained and any surgery in this patient population should be preceded by a full discussion and assessment among the surgeon, hematologist, anesthetist, and the patient. The clinical team must decide whether to accept the constraints of bloodless surgery and decide whether the procedure for the JW patient has an acceptable mortality risk. Preferably, the surgeon should be experienced in treating JW patients or be familiar with bloodless surgical techniques; if not adequately trained or experienced, the surgeon may choose to refer the JW patient for consultation.[10-12] Once the full implications of the risks and benefits of the procedure and of alternative treatments are discussed, the preoperative planning should be made.

Furthermore, this principle which is applied in JWs should be implemented for all patients undergoing cardiac surgery. Bloodless surgery should not be limited to JWs, instead it should form an integral part of everyday surgical practice.[10,13] More importantly, patient's consent is obtained for blood transfusion requirements in the emergency setting, in which the person called the JW is brought and he/she is often unconscious. In such cases where the place of the will is regarded as legal, the term default consent is used. Regarding to the principle of 'primum non nocere', the treating physician should demonstrate every effort to save the life of an unconscious patient ignoring his/her wishes, even if the physician is aware of that the patient refuses blood transfusion.[4] In this case, aforementioned bloodless surgical strategies were successfully performed and the patient remained without having a blood transfusion need and his recent hemoglobin value was lower than normal.

In conclusion, we suggest that it is of utmost importance to make essential medical attention to such a case of an aortic fibroelastoma with a coronary lesion, even he is a Jehovah's Witnesses, being aware of the physician's ethical and legal responsibilities and applying the main principle of respect: Primum non nocere.

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