# MALFORMATION SYNDROME WITH TOTAL SITUS VISCERUM INVERSUS - CONTRAINDICATION FOR TISSUE DONATION

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## Abstract

We report a case of a 65-year-old woman who died with evidence of acute cardiac ischemia, despite receiving emergency medical care. The woman's body was promptly brought for autopsy to the Department of Forensic medicine and deontology, Medical University-Sofia, Bulgaria and placed in a forensic storage refrigerator. During the immediate external examination of the body and based on the results of the performed serological tests of the blood samples taken from the deceased, no medical data for contraindication for tissue donation were established. A conversation was made by the hospital donation coordinator with the daughter of the deceased for the possibility of tissue donation, where she gave her consent. In principle, to perform such a procedure in the Department of Forensic medicine and deontology, an autopsy should always be performed to exclude contraindications for tissue donation, and after the autopsy the standard documents required for this purpose are signed by the relatives. The autopsy of the deceased revealed the presence of a total situs viscerum inversus, as well as ischemic heart disease with multiple disseminated myocardiofibrosis, acute anterior septal myocardial infarction, two atrial septal defects and severe concentric hypertrophy of the heart. According to standard medical criteria, all types of genetic malformations are a contraindication for donation, which is why, in this case, the donation procedure was terminated.

***Key words:*** *malformation syndrome, situs viscerum inversus totalis, tissue donation, contraindications*

## Introduction

Organ and tissue donation occurs when organs and tissues are recovered from a person who has recently died [1]. Transplantation is a very successful way of saving and improving the lives of people who are experiencing serious health problems. Every person may be considered a potential donor [2]. Donated tissue such as skin, bone, ligaments and heart valves can be used in many surgical applications and life-threatening medical situations including saving patients with severe burns, allowing athletes with torn ligaments or tendons to heal and regain strength, restoring hope and mobility to military men and women who have been injured in combat, and repairing musculoskeletal structures such as teeth, skin, and spinal components. This can dramatically improve the quality of life for recipients, and help save lives. One tissue donor can heal the lives of more than 75 people [1, 2, 3].

Health professionals play a vital role in the tissue donation process by identifying and referring potential donors [2, 3, 4]. In the department of Forensic medicine and deontology, Medical University-Sofia, the Tissue Bank coordinator reviews the data from the performed autopsy and any other relevant medical information, determine whether the potential tissue donor

is eligible to donate, and approaches the family with the option of donation. One of the most important steps in this process is to exclude any contraindications for tissue donation [5]. Absolute and major contraindications in Bulgaria for tissue donation are: age greater than 80 years for men and 75 for women; Lab diagnosed infections (e.g. MRSA, VRE or C. difficile); Blood cancers (e.g. Lymphoma, Leukemia, Myeloma) and other types of cancers; Neurological diseases (e.g. ALS, MS, Alzheimer's, Parkinson's, Dementia); HIV, Hepatitis B, Hepatitis C, HTLV I/II, active TB; Severe Sepsis; Genetic malformations, etc.

**Case presentations:** We report a case of a 65-year-old woman who died with evidence of acute cardiac ischemia, despite receiving emergency medical care. The woman's body was promptly brought for autopsy to the Department of Forensic medicine and deontology, Medical University-Sofia, Bulgaria and placed in a forensic storage refrigerator. During the immediate external examination of the body and based on the results of the performed serological tests of the blood samples taken from the deceased, no medical data for contraindication for tissue donation were established. A conversation was made by the hospital donation coordinator with the daughter of the deceased for the possibility of tissue donation, where she gave her consent. In principle to perform such a procedure in the Department of Forensic medicine and deontology, an autopsy should always be performed to exclude contraindications for tissue donation, and after the autopsy the standard documents required for this purpose are signed by the relatives.

The autopsy of the deceased revealed the presence of a complete "mirror" transposition of the internal organs, a condition called "total situs viscerum inversus". (Photo 1, photo 2)

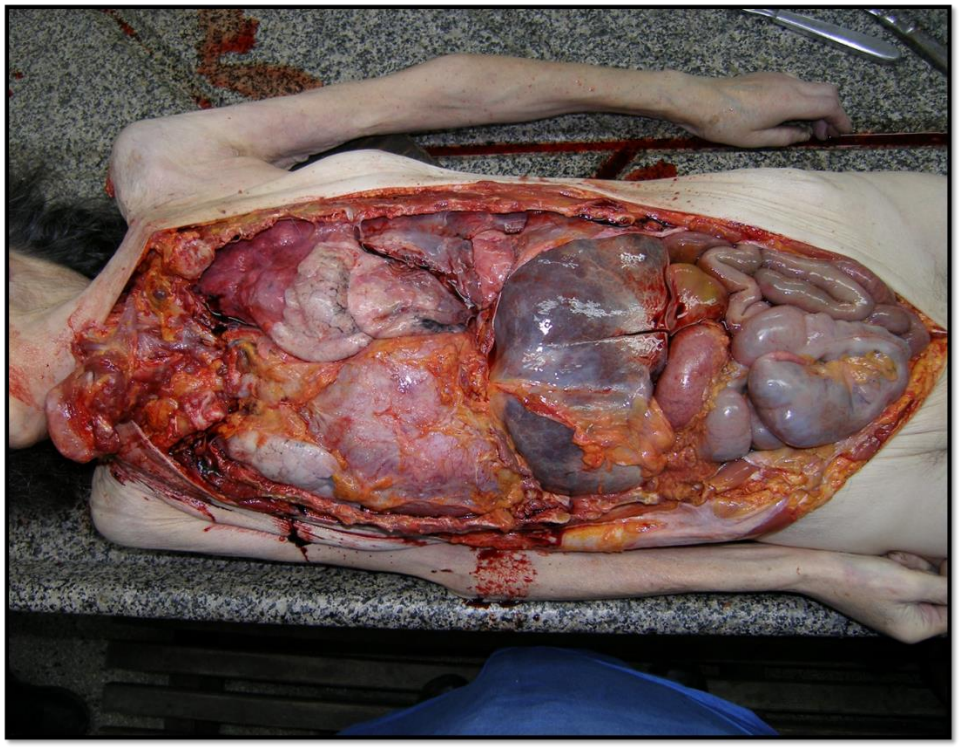

Photo 1. Total situs viscerum inversus.

Photo 2. Total situs viscerum inversus – evisceration.

The lungs filled the thoracic cavity; they were heavy, with a grayish-purple color and a smooth surface. In addition, their cut surface was purple-bluish in color, from it spontaneously and under lateral pressure, pink foamy liquid flowed abundantly. The heart was greatly enlarged at the expense of all cavities. The chamber from which the aorta emerged was located on the right with 30 mm thickness of the wall. The chamber from which the pulmonary artery emerges was located on the left and was 10 mm thick. The aorta passed in front of the pulmonary artery and descended to the right of the spine. Both atria were 6-7 mm thick. The mitral valve had fibrous changes. The aortic valve has no visible changes. The tricuspid and pulmonary valves have an enlarged ring. Two defects in the septum of the atriums were observed, one with a diameter of 30 mm and the other with a diameter of 7 mm (Photo 2).

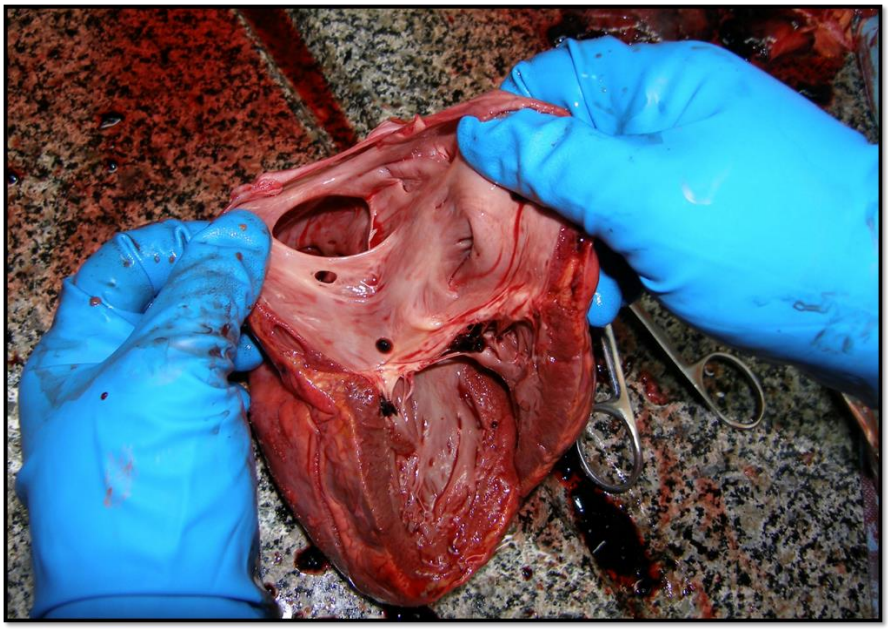

Photo 2. Atrial septal defects.

Furthermore, ischemic heart disease with multiple disseminated myocardiofibrosis was present with acute anterior septal myocardial infarction, and severe concentric hypertrophy of the heart. The organs in the abdominal cavity have a "mirror" position as well.

**Discussion and conclusion**

Situs inversus totalis is a rare, genetic, developmental defect during embryogenesis characterized by total mirror-image transposition of both thoracic and abdominal viscera across the left-right axis of the body [6, 7]. Congenital abnormalities, such as primary ciliary dyskinesia [8], Kartagener type, polysplenia syndrome, biliary atresia, congenital heart disease, and midgut malrotation, as well as vascular anomalies (e.g. absence of retrohepatic inferior vena cava, preduodenal portal vein, aberrant hepatic arterial anatomy) and malignancy, are frequently associated.

The genetics of situs inversus is complex. Several familial cases have been reported in which the inheritance has been described as either autosomal recessive (most commonly), autosomal dominant, or X-linked [9]. The condition appears to be genetically heterogeneous, meaning that different genetic factors or genes may cause the condition among different people or families [6, 10]. If situs inversus is associated with another underlying syndrome or condition, the inheritance pattern may be the same as that of the underlying condition.

According to standard medical criteria, all types of genetic malformations are a contraindications for donation, which is why, in this case, the donation procedure was terminated.

Statement for Potential Conflicts of Interest: There is no potential conflict of interest.

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