

PATHOLOGY

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THORACO-OMPHALOPAGUS CONJOINED TWINS (case report)

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Abstract: Conjoined twins develop due to impairment in embryo division process. There are some risk factors considered to increase the incidence of this condition. However, it is necessary to perform further investigation of genetic processes as well as teratogenic agents involved. In this study we review a case of an 18-year old patient with the first pregnancy, having no apparent teratogenic factors or family history, diagnosed with conjoined twins. An abortion was performed on the 13th-14th weeks of gestation. Multiple malformations of conjoined twins were studied and genetic counseling was prescribed to the patient during planning of the next pregnancy.

KeyWords: conjoined twins, thoraco-omphalopagus twins, malformations.



INTRODUCTION

Conjoined twins are monozygotic twins, formed as a result of incomplete division of the embryo, having common extraembryonic organs: chorion, amnion, placenta. The incidence of this condition ranges from 1:50,000 to 1:200,000 births [1, 2]. Understanding exact mechanisms triggering the formation of conjoined twins requires further investigation. It is assumed that the late embryo division, which occurs on days 13-15 of the development, is the leading reason of non-separation of the twins. The conjoined twin formation is associated with the development of anomalies of the twins and a high perinatal mortality rate. Considering the above mentioned, the study assessed the role of ultrasound diagnosis in the management of conjoined twin pregnancies.

CASE STUDY

An 18-year-old patient K. at 13th-14th week of gestation of the first pregnancy was referred for planned ultrasonography. Ultrasonography showed multiple fetal malformations. She was found to have conjoined twins, hypoplasia of both fetal nasal bones, cystic hygroma in the occipital region of one fetus. The bones of the pelvis and lower extremities of one fetus were not detected. An abortion for medical reasons was recommended and the patient was admitted to Odessa Regional Clinical Hospital. According to the collected data, the course of pregnancy was normal and there were no cases of hereditary diseases in the family history. An abortion was performed and the abortion material was sent to Odessa Regional Pathologic Bureau. The fetuses were labeled as fetus No.1 and fetus No.2 and referred to autopsy. The fetuses were joined in the chest and abdomen areas (Terata Anacatadidyma) (Fig. 1).

Fetus No.1. The brain (3.5 x 2.5 x 2 cm) was formed according to the stage of the development, lateral ventricles were slightly expanded and meninges were plethoric. The heart (1.2 x 0.7 x 0.7 cm) was visualized under the sternum following the dissection of the chest. The heart had a normal structure with only one vessel leaving it (aorta) and was not covered by pericardium. The esophagus and the

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trachea were without abnormalities. The spleen and kidneys were located at normal anatomical sites.

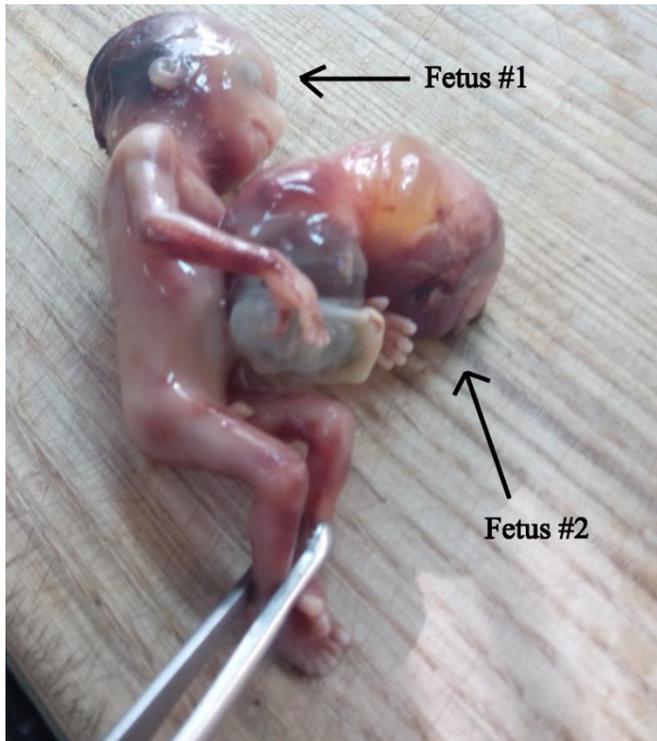


Figure 1. The fetuses joined in the chest and abdomen were labeled fetus No.1 and fetus No.2.

Fetuses had one common abdomen (Fig. 2). The abdominal cavity was found to have common intestines and common liver (spherical form, 2.5 x 2 x 1.5 cm) with a gallbladder.

Fetus No.2. There was severe edema and a closed cavity (4 x 2.5 x 1 cm) filled with transparent, slightly yellowish contents in the occipital area. The neck was drastically shortened. The fetus was found to have humerus bud. The brain (2.5 x 2.3 x 3.2 cm) was formed according to the stage of the development, meninges were plethoric. The heart (0.9 x 0.5 x 0.5 cm) had cervical ectopy and was not covered by pericardium and with only two chambers inside.

The examination determined lung aplasia.

The bones of the pelvis and lower extremities were not detected.

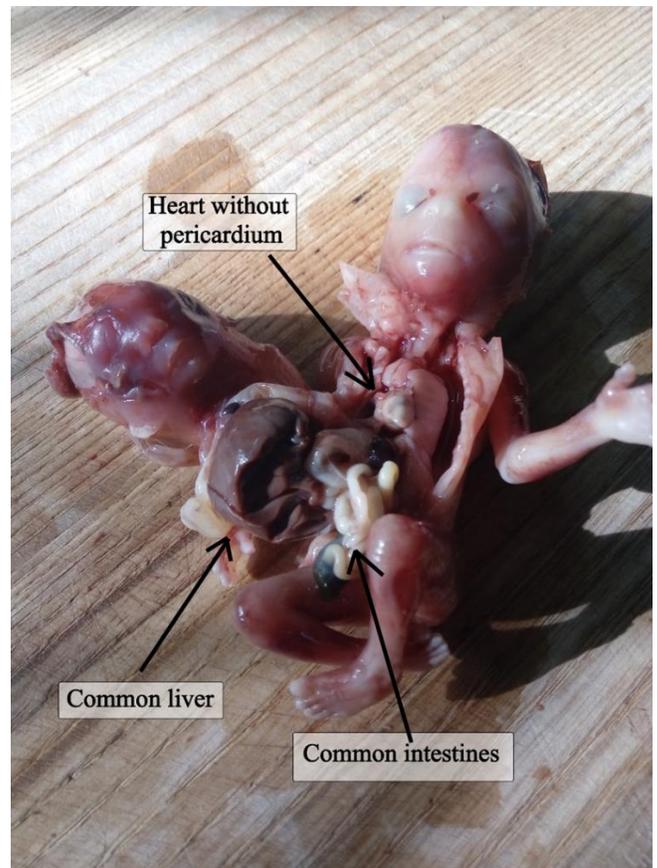


Figure 2. Common abdomen in fetuses. Common intestines and common liver with a gallbladder in the abdominal cavity.

The common liver was connected with the heart through the blood vessel (Fig. 3).

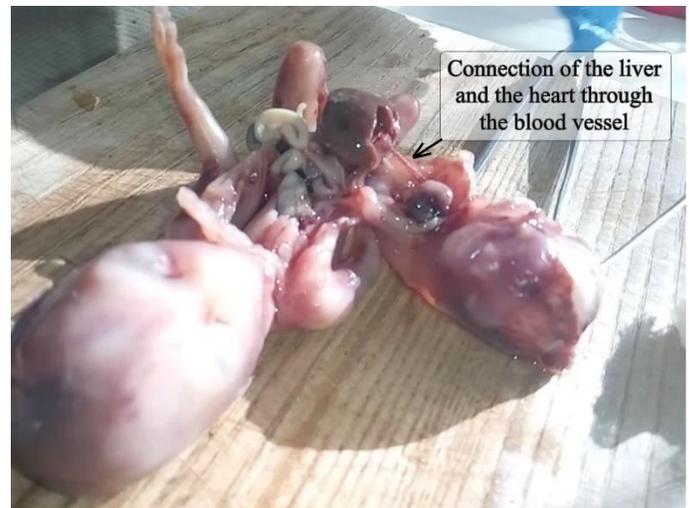


Figure 3. Common liver connected with the heart of fetus No.2 through the blood vessel.

The autopsy findings were confirmed by investigation of histological material.

DISCUSSION

After the fertilization, a zygote is going through a number of stages resulting in a formation of the fetus. Sometimes a zygote or an embryo divides, resulting in the formation of two fetuses. This process is very rare (6-9 cases per one thousand births during natural pregnancy [3]). Normally in monozygotic twins, an embryo division occurs from the 2nd to the 7th days of the development, leading to the formation of two separate fetuses. Sometimes, under uncertain mechanisms, this process can be prolonged up to 13-15 days and terminated with an incomplete division. In this case, both fetuses have common extraembryonic organs and are joined in different parts of their bodies, or even sharing common organs [4]. In some clinical cases it is possible to perform surgical separation, but it entails additional risks and can be life-threatening to one or both of the twins, especially when the surgery affects vital organs [5]. Sometimes, as in the present clinical case, surgery is impossible or it is of no use due to multiple malformations of the fetuses [6]. Etiology of the formation of conjoined twin is still uncertain, but some assumptions can be made: for example, there are authors indicating assisted reproductive technologies as a risk factor. Some studies showed that 14.8% of 75 pregnancies with conjoined twins were observed in pregnancies after artificial fertilization [7]. Such techniques as ovulation induction, intracytoplasmic sperm injection and assisted hatching may play a significant role in the failure of an early embryo division [8] but the amount of such investigations is considered not sufficient. In both natural pregnancy and assisted reproduction, some teratogenic agents (griseofulvin [9] or ionizing radiation, as in the case of Chornobyl-impacted regions [10]) and genetic impairments [1, 11] significantly increase the risk of the formation of conjoined twins, but no unified point of view has been suggested at this point. Unexplained etiology and mechanisms of formation of conjoined twins determine the necessity of thorough management of physiological pregnancy and increase the role of ultrasound diagnosis, which can detect formation of conjoined twins as early as at 7-9th weeks of gestation [12].

4 CONCLUSIONS

Couples with conjoined twins in family history should undergo genetic counseling to consider all possible risk factors.

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