Siamese twins – prenatal diagnosis in the first trimester of pregnancy. Case study and review

Bliźnięta syjamskie – diagnoza prenatalna I trymestru ciąży. Analiza przypadku oraz przegląd literatury

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Abstract

Conjoined twins are a unique type of monozygotic twins. All monozygotic twins should be thoroughly evaluated for incomplete separation and, if incomplete separation is diagnosed, the extent of internal organ connection and the presence of additional developmental anomalies of the foetuses should be assessed. Common heart of foetuses is particularly difficult to diagnose and crucial for prognosis.

We present an example of female thoracoomohalopagus twins with a common triventricular heart, connate livers, and joined hepatic vessels, diagnosed in week 12 of pregnancy. Due to the high complexity of foetal connection, separation was not possible and following interdisciplinary consultation, the pregnancy was aborted upon the patient's request in week 16.

Key words: **conjoined twins** / **thoracoomphalopagus** / **single heart** / **ultrasonography** /

Streszczenie

Bliźnięta nierozdzielone stanowią unikalny typ bliźniąt monozygotycznych. Wszystkie bliźnięta monozygotyczne winny być wnikliwie ocenione w kontekście ich niecałkowitego rozdzielenia, a w sytuacji postawienia takiej diagnozy należy ustalić rozległość zespolenia narządów wewnętrznych oraz obecność dodatkowych anomalii rozwoju płodów. Szczególnie trudną diagnostycznie oraz kluczową w kategoriach rokowania jest sytuacja wystąpienia wspólnego serca płodów.

Prezentujemy przypadek bliźniąt płci żeńskiej typu thoracoomphalopagus, ze wspólnym, trójkomorowym sercem, zrośniętymi wątrobami oraz połączonymi naczyniami wątrobowymi rozpoznany w 12 tygodniu ciąży. Z powodu wysokiego stopnia złożoności połączenia płodów operacja rozdzielnia bliźniąt nie była możliwa i po interdyscyplinarnej konsultacji ciąża została na prośbę pacjentki zakończona terminacją w 16 tygodniu.

Słowa kluczowe: **bliźnięta syjamskie** / **thoracoomphalopagus** / **pojedyncze serce** / / **ultrasonografia** /

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Introduction

Conjoined twins are a rare type of monozygotic twins. They occur in 1:50 000 to 1:100 000 births and are three times more frequent in female foetuses [1, 2]. An attempt to explain this phenomenon is based on a theory that two processes coincide: the monozygote divides and X chromosome is inactivated. It is not known how conjoined twins develop. No karyotype variations that could decide on the occurrence of such defect have been found so far. There are also no studies proving any predispositions related to race, geographical location, family history of conjoined twins, the method of impregnation, or relatedness of parents [3].

At present, two theories that could explain the process of conjoined twins' development are known. The first theory, the theory of fission, assumes that the germinal disc undergoes incomplete separation between day 13 and 15 following impregnation. The other theory, called the theory of fusions, assumes that the impregnated ovum divides completely, but they are joined again due to the similarity of stem cells in both embryos. The theory of fusion is believed to be more probable [4]. Conjoined twins share the amnion, chorion, and placenta; such a situation, however, is not characteristic of conjoined twins, as it occurs in 1% of monozygotic twins [5].

According to the classification proposed by Spencer et al., there are seven types of conjoined twins. The names of the individual types are formed based on the anatomical structure constituting the largest part of connection. The suffix – pagus, of Greek origin, means 'tied, joined' [6].

The following groups of conjoined twins have been distinguished:

thoracopagus – connection at the level of the chest,
omphalopagus – connection at the level of epigastrium,
pygopagus – fusion of sacral bones,
ischiopagus – connection of the ischia,
craniopagus – connected skulls,
parapagus – extensive connection of the torso,
rachipagus – connection along the spine.

The most frequent type of all conjoined twins is thoracopagus (40%) and the rarest is craniopagus [7]. Mixed types are also found, like the discussed case – thoracoomphalopagus – that combines features of two groups. Presently, ultrasound scans play the most important role in diagnosing conjoined twins. Early diagnosis is crucial, as it allows for evaluating the type of conjoining, determining a prognosis, planning the conduct during pregnancy, preparing delivery, and prenatal treatment, or taking the decision to terminate the pregnancy. It is also important to diagnose concomitant defects such as defects of the neural tube and facial skeleton, imperforate anus, diaphragmatic hernia, or heart defects.

Conjoined twins can be diagnosed using transvaginal ultrasound scans early in pregnancy. A case of very early diagnosis, already in week 7 + 6 days has been described [8]. Usually, however, such diagnosis is made during the screening prenatal examination in the first trimester.

Case description

A 31-year-old patient came for prenatal examination in week 12 of her pregnancy. Referred as second pregnancy following natural impregnation, with no history of miscarriage. Ultrasound scanning revealed pathological twin pregnancy: conjoined twins, thoracoomphalopagus, with single umbilical cord, and the chorion located on the anterior wall. The volume of amniotic fluid was normal. Nuchal translucency of 1.7 mm and 1.2 mm was determined in the examination of foetuses.

The patient was referred to a referral centre, in which the initial diagnosis was confirmed in week 14 during an ultrasound scan. Moreover, nuchal translucency progressed in one of the twins (CRL 81 mm, NT 3.1 mm), while the initial value did not change in the other (NT 1.7 mm). Additional echocardiography was performed in the foetus, in which triventricular undivided heart was found. The aortic arches of both twins were visible, while pulmonary arteries could not be assessed in ultrasound scanning. Superior and inferior vena cava connected to the right atrium were seen in one of the twins. Two venous ducts supplying both foetuses were also found. Female genitourinary organs were visible.

Ultrasound assessment and interdisciplinary consultation led to the conclusion that the twins could not be separated after birth. This information was given to the mother of the conjoined twins, who decided to terminate the pregnancy; termination was done in week 16 of pregnancy with the use of misoprostol, a synthetic prostaglandin E1 analogue. Autopsy performed confirmed the diagnosis.

Discussion

The occurrence of a developmental defect in the form of conjoined twins is associated with many complications that can lead to intrauterine demise of twins due to the death of one of the foetuses or the coexistence of lethal defects. In foetuses that survive, the possibility of difficulties during delivery should be taken into account; such difficulties can be due to the size of the conjoined foetuses and their position in relation to each other. Such pregnancies are usually delivered through a C-section in week 36-38 of pregnancy, but reports of natural births also exist [9].

The most important element deciding about the condition of conjoined twins and their prognosis is the presence of shared organs or the fusion of vital system parts. The condition of each of the twins, ie additional systemic defects or chromosomal aberrations, is another prognostic factor. In the case of conjoined twins, the condition of one twin significantly influences the condition of the other.

Thoracoomphalopagus twins are connected at the level of the thorax and the upper part of the abdominal cavity. Apart from a shared sternum, diaphragm, and usually pericardial sac, various combinations of severe anomalies of the circulatory system and of liver anatomy are often observed. In 25% of cases, a shared hepatic lobe occurs together with abnormal bile ducts and hepatic vessels. In 50% of cases, the small intestine can be connected in the duodenal section [10].

The prevalence of heart defects in twins conjoined at the level of the thorax is very high. In 90% of cases, the pericardium is fused and the connections of superior mediastinal vessels are pathological in 75% of foetuses [7].

Anatomical anomalies of conjoined twins' hearts have been grouped into 4 categories:

group A: separate hearts, separate pericardia,
group B: separate hearts, joined pericardium,
group C: fused atria, separate ventricles,
group D: fused atria, joined ventricles.

Defects from group D are characteristic of thoracopagus [7]. The anomaly found in the presented case belongs to group C.

The assessment of heart fusion and the diagnosis of comorbid heart defects is extremely significant in the qualification for surgical separation of twins in the postnatal period, but it is also a prognostic factor deciding about long-term survival. Echocardiography and electrocardiography are performed to determine anomaly characteristics in the case of fused hearts and to assess the fusion surface in these organs. These tests are also used to observe abnormalities in heart function physiology. The presence of a single heart function signal in ECG usually indicated that the separation of twins will not be possible. Two separate recordings give better prognosis [1]. The complexity of cardiac connections in conjoined twins can be very high and the assessment of single heart haemodynamics requires advanced ultrasound machines and a very experienced physician performing the examination. In advanced pregnancies (2nd and 3rd trimester) the obligatory ultrasound scan should be followed up with an MRI.

Prenatal diagnostics of conjoined twins is to provide all obtainable information enabling the evaluation of survival odds for the foetuses and the assessment whether twins can be separated after birth. A single heart is a particularly difficult challenge for surgeons. In the last 10 years, the pubmed.com scientific database noted only 11 cases of successful separation of twins with a single heart.

The diagnosis of conjoined twins poses many ethical dilemmas. Questions arise: can the twins be separated and should separation be attempted despite the lack of significant, rational chance of survival? What will be the quality of life of the twins after separation? Will both of them live? Should the twins be separated if only one of them will survive? Should pregnancy be terminated if the diagnosis is known and separation will not be possible? The final decision to continue or terminate pregnancy is made by the parents once they obtain accurate information from a team of physicians.

The general death rate of conjoined twins is high, at about 85%. It is difficult to assess it due to a high intrauterine death percentage (about 60%) [11] and the option of pregnancy termination [12].

A decision on the further life of foetuses and the planning of the best postnatal conduct is possible only on the basis of early prenatal diagnosis with a detailed evaluation of joining extent in conjoined twins. The basic tool for obtaining the necessary information is ultrasonography. In the described case, the thoracoomphalopagus defect was diagnosed during the first prenatal examination, hence early multidisciplinary and multicenter consultation was possible; as a result, the final diagnosis could be made and prognosis could be determined.

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Piśmiennictwo

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