



CASE REPORT / ПРИКАЗ БОЛЕСНИКА

Conjoined twins – *parapagus dithoracicus*

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SUMMARY

Introduction Throughout history, various types of conjoined twins have been described, many of whom lived until adulthood. The anomaly includes several varieties, with parapagus being one of the rarer forms of conjoined twins.

The aim of this paper was to present a rare case of conjoined twins – parapagus type.

Case outline In this paper we present autopsy and radiologic findings in male 40-week-old parapagus twins from a monozygotic pregnancy, with multiple anomalies in different systems of organs and musculo-skeletal systems. Parapagus twins have a side-to-side connection with shared pelvis and can be defined as one of three subtypes. In our case, it was a dithoracic parapagus subtype, with four upper and three lower limbs (*tetrabrachius tripus*), where the presence of the third bizarre-looking lower limb made the case even more unusual among the rare type. Both parents were drug addicts with a history of sedative abuse. The mother had regular sonographic examinations during pregnancy.

Conclusion Adequate early prenatal diagnosis of conjoined twins and possible termination of such a pregnancy are the key reasons for rare occurrence of such cases. Failure to recognize a pathological pregnancy and its maintenance until the delivery, despite regular gynecological examinations, is why our case is unique in comparison to contemporary literature.

Keyword: conjoined twins; dithoracic parapagus; autopsy

INTRODUCTION

Conjoined twins (Siamese twins) occur during twin pregnancy in which identical fetuses are connected with certain parts of the body and share certain organs, with monoamniotic and monochorionic placenta [1]. The frequency of anomaly is 1:50,000–100,000 births, where only 18% of twins are newborns, since most of conjoined twin pregnancies result in still births and miscarriages [1, 2]. Etiopathogenesis is still unknown, but it is thought that the primary event occurs 13–15 days after fertilization, where, due to incomplete fission (i.e. separation of a zygote) or due to fusion of two embryos, the formation of a physical connection between the fetuses is made [3]. The theory of incomplete splitting is a commonly held scientific opinion [4, 5, 6].

The name 'pagus' comes from the Greek word *pagus*, meaning 'connections', and points to the connection between two fetuses, so the classification of conjoined twins is based on the anatomic characteristics of that connection [7, 8]. The most common are thoraco-omphalopagus (28.5%), thoracopagus (18.5%), omphalopagus (10%), and parasitic twins (10%). Other types, including parapagus, are much rarer [3–9].

Parapagus twins are connected laterally and have a single pelvis with a shared pubic symphysis, while the organs may or may not be

shared [10]. They are classified as dithoracic parapagus (two heads with two thoraces), dicephalic parapagus (two heads connected to a single chest), and diprosopus parapagus (two faces on one head) [11].

In our paper we present a case of male parapagus twins, type dithoracic parapagus, with four upper and three lower extremities (*tetrabrachius tripus*), born despite regular gynecological examinations of the mother, with autopsy findings.

CASE REPORT

A 20-year-old primigravida with gestational age of 40 weeks was admitted to the regional hospital as an emergency case with abdominal pain. The heteroanamnesis data collected from the closest relatives indicated that the pregnancy on a regular ultrasound examination was diagnosed as a normal twin pregnancy. Both parents were drug addicts with a history of sedative abuse and at the time of birth the father was hospitalized on suspicion of a tuberculous process.

On admission, the patient was examined by a gynecologist and an anesthesiologist. Ninety minutes after admission, the pregnancy was ended by cesarean section. Cardiotocography

Received • Примљено:

October 3, 2017

Revised • Ревизија:

January 10, 2018

Accepted • Прихваћено:

January 15, 2018

Online first: January 30, 2018

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Figure 1. Posterior (a) and anterior (b) view of parapagus twin males



Figure 2. A rudimentary lower limb

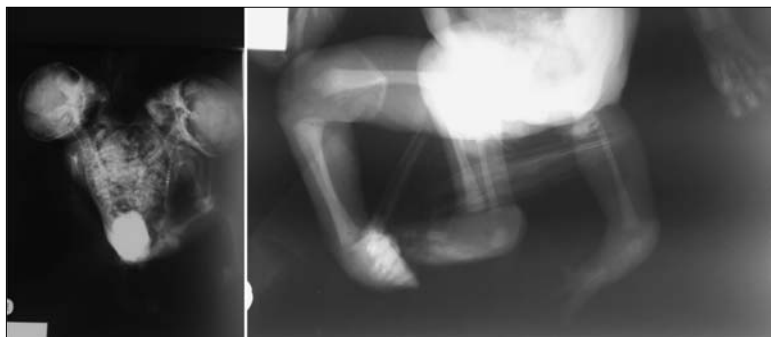


Figure 3. Postmortem X-ray shows two separate spines and visible third limb with one bone



Figure 4. Opened thoracic cavity showing two separate hearts

record done 60 minutes before birth indicated a normal heartbeat. While extracting the twins, it was found that they were conjoined. The Apgar score one minute after birth was 2; fatal outcome occurred several minutes after birth. The autopsy was performed in two acts – initially when the head and chest were open, and the continuation of the autopsy took place later, after fixating the tissues and organs in formalin. We considered that the fixation would improve the visibility of morphological and anatomical relationships between organs and vascular structures.

The twins were fused anterolaterally, from mid-chest to the navel, with a single pelvis (Figure 1). Crown–heel length of twin A was 47 cm, the crown–rump length was 30 cm; head circumference in both twins was 31.5 cm. Common chest circumference was 38 cm, and the combined weight was 4.2 kg at birth. Fetus A had normally developed two upper and two lower limbs, with no anomalies of the fingers. Both feet were in the equinovarus position. The upper extremities of fetus B were normally developed, but there was one bizarre-looking rudimentary lower limb, with elongated feet and ectrodactyly and syndactylic fingers. Three unusual fingers were presented with creations in place of nail plates that had the appearance of claws (Figure 2). A postmortem-made X-ray of the twins indicated the existence of two separate spinal columns and one common pelvis (Figure 3). Lanugo hairs were present; nails were at the level of the fingertips. Both fetuses had external genitalia of the male type, with two testicles in twin A and one testicle in the scrotal bag of twin B. There was one anus.

The placenta of the twins had one amnion and one chorion, one umbilical cord with three blood vessels – one vein and two arteries. Autopsy findings revealed that the fetuses had a shared sternum, but two separate thoracic cavities, where fetus A had a pronounced defect in the

diaphragm with the penetration of the liver and intestines into the chest cavity, with severe hypoplasia of both lungs, while fetus B had a normally developed diaphragm. In the thoracic cavity of the fetuses, fully developed hearts without any anomalies were present, between which there was no communication (Figure 4). Both hearts were in one common pericardial sack. Each fetus had an aorta with an anatomically appropriate origin and the normal anatomical disposition of associated blood vessels. It was found that fetus A had a usual morphology of the thoracic and abdominal aorta and its branches, while the aorta of fetus B flowed in the abdominal part of the aorta of fetus A.

The twins had a common abdominal cavity, with a separately developed esophagus and stomach of each twin. There were two separated, parallel to each other, small intestines. At 20 cm before the ileocecal valve, small intestines were connected, where they continued as one shared, forming the remaining part of the small intestine and the colon, with no anomalies (Figure 5).

Fetal livers were separate in each, where the liver of fetus A was placed in the thoracic cavity because of the diaphragm defect. Both fetuses had a pancreas in appropriate localization. The urinary system was found only in twin A – two apparently normally developed kidneys and adrenal glands, with two ureters, the urinary bladder, and the urethra.

DISCUSSION

Conjoined twins are a rare medical phenomenon. It is believed that the frequency of the occurrence is 1:200 of monozygotic twin pregnancies [12]. Among the first cases described in the literature were sisters Mary and Eliza



Figure 5. Place of conjunction of two parallel small intestines and separate stomachs

Chulkhurst from England, the so-called Biddenden Maids, which were joined at the shoulder and the hip, and the case of Chang and Eng Bunker, born in 1811 in Siam, today Thailand, after whom this anomaly is named “Siamese twins” [2, 13, 14].

The most common forms of conjoined twins are thoracopagus and thoraco-omphalopagus, while the parapagus incidence is lower [7, 15, 16]. It was noted that parapagus more often develops in male twins, compared to other varieties of Siamese twins, which are generally more common in female fetuses [2]. The etiology of the disorder is not known yet, but conjoined twins, with no statistically significant differences, can develop in a primigravida or in a multigravida, and it is shown that maternal age is not related to this type of twin pregnancy [17]. In our case, the mother was a 20-year-old primigravida, while both parents had a history of abuse of narcotics and sedatives.

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The information on the type of narcotics and sedatives, as well as on the period when they were used, was missing. In the literature, there is still no information on the possible risk factors, other than the method of assisted reproduction, whose impact is still not fully understood, but in our case, the question arises whether the abuse of opiates and sedatives had any influence on the development of the malformed twins [18].

Autopsy findings showed that the twins’ hearts had no anomalies, but both hearts were found in the common pericardial sack [8, 19]. Extremely rare within the parapagus is the existence of diaphragmatic hernia with severe hypoplasia of both lungs, as found in our case. Therefore, death of fetus A occurred very soon after birth, when normal respiratory function should be established, and considering that the vascular system of fetus B was anatomically and functionally linked to fetus A, death of fetus B occurred as a result of fetus A dying. In addition, both fetuses had functionally shared kidneys. Other abnormalities that can be seen in the parapagus type of conjoined twins are the common lower part of the gastrointestinal system and abnormalities of the feet, which were present in our twins [20, 21]. The third bizarre-looking lower extremity that belonged to fetus B had an anomaly known as distal phocomelia [22].

Early prenatal diagnosis of conjoined twins is of great importance. The literature states that from the 12th week of gestation, the diagnosis of conjoined twins pregnancy can be made [18, 23, 24]. Adequate early prenatal diagnosis and possible termination of such a pregnancy are the key reasons for the rare occurrence of such conditions. Early diagnosis is essential for parents, but also for planning adequate postnatal care. However, in the presented case of male parapagus twins, both parents stated that the mother had regular gynecological examinations and regular ultrasound scans, approximately once a month, which failed to detect numerous anomalies and, therefore, up until the delivery, the pregnancy was considered a normal twin pregnancy. The pregnancy ended with a cesarean section because the gynecologists thought that the fetuses were not in the adequate position for labor. Only at birth was it found that the twins were conjoined. A failure to recognize a pathological pregnancy and its maintenance until the delivery, despite regular gynecological examinations, is why our case is unique in comparison to the contemporary literature.

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Сијамски близанци – *parapagus dithoracicus*

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САЖЕТАК

Увод Описано је више врста спојених близанаца, од којих су многи доживели одрасло доба. Сијамски близанци обухватају неколико варијетета, од којих је парапагус форма међу ређима.

Циљ овог рада је био да прикажемо редак случај сијамских близанаца парапагус типа.

Приказ болесника Приказани су обдукциони и радиолошки налази мушких спојених близанаца парапагус типа старих 40 недеља, са вишеструким аномалијама на различитим органским системима и мускулоскелетном систему. Парапагус тип се карактерише бочним спојем два близанца, са једном, заједничком карлицом и може се класификовати у три подтипа.

У нашем случају радио се о *parapagus dithoracicus* субтипу, са четири руке и три ноге (*tetrabrachius tripus*), где је присуство трећег екстремитета бизарног изгледа учинило случај још ређим варијететом. Оба родитеља су зависници од опојних средстава са злоупотребом седатива. Током трудноће мајка је уредно контролисана редовним ултразвучним прегледима. **Закључак** Адекватна пренатална дијагноза сијамских близанаца и могући прекид такве трудноће су кључни разлози за релативно ретку појаву оваквих догађаја. Непрепознавање патолошке трудноће и њено одржавање, и поред редовних сонографских прегледа, оно је што овај случај чини јединственим.

Кључне речи: сијамски близанци; парапагус тип; обдукција