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Case Report / Приказ случаја

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Conjoined twins: Parapagus Dithoracicus

Сијамски близанци: *Parapagus dithoracicus*

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SUMMARY

Introduction Through the history there have been described various types of conjoined twins, many of whom lived until adulthood. Anomaly includes several varieties, with parapagus being one of rarer form of conjoined twin.

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

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

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

Keyword: conjoined twins; parapagus dithoracic; autopsy

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 случаја Приказани су обдукциони и радиолошки налази мушког парапагус типа спојених близанаца 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 је оно што овај случај чини јединственим.

Кључне речи: сијамски близанци; парапагус тип: обдукција

INTRODUCTION

Conjoined twins (siamese twins) occur during twin pregnancy in which identical fetuses are connected with a 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 [2], where the only 18% of twins are newborns, since most of conjoined twins pregnancies result in still births and miscarriages [1]. Etiopathogenesis is still unknown [3], but is considered that the primary event occurs between 13 and 15 days after fertilization, where due to incomplete fission (i.e. separation of a zygote) or due to the fusion of two embryos the formation of a physical connection between the fetus is made. The theory about incomplete splitting is a commonly held scientific opinion. [4, 5, 6].

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

Parapagus twins are connected laterally and have single pelvis with a shared pubic symphysis, while the may or may not be shared [10]. They are classified as: *dithoracic parapagus* (two heads with two thorax), *dicephalic parapagus* (two heads connected to a single chest) and *diprosopus parapagus* (two faces on one head) [11].

In our paper we present the case of male parapagus, type *dithoracic parapagus*, with four upper and three lower extremity (*tetrabrachius tripus*), born despite regularly gynecological examinations of the pregnant woman, with autopsy findings.

CASE REPORT

A young 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 that was collected from the closest relatives indicated that the pregnancy on a regular ultrasound examination was diagnosed as usual twin pregnancy. Both of the parents were drug addicts with the abuse of sedatives and at the time of the birth the father was hospitalized on suspicion of tuberculous process.

On admission patient was examined by a gynecologist and anesthesiologist. An hour and a half after admission the pregnancy was ended by cesarean section. Cardiotocography record that was done 60 minute before birth indicated a normal heartbeat. While extracting the twins it was found that they were conjoined twins. The Apgar score in 1st minute was 2; a fatal outcome occurred a few minutes after birth. An autopsy was performed in two acts, first when the head and chest were open, and a continuation of the autopsy was followed later, after fixation of tissues and organs in formalin. We considered that the fixation would improve the visibility of the morphological and anatomical relationships between organs and vascular structures.

The twins were fused anterolateral, from mid-chest to the navel with a single pelvis (Figure 1). Crown-heel length of twin A was 47 cm, a crown-rump length was 30 cm; head circumference in both twins was 31.5 cm. Common chest circumference was 38 cm, and a combined weight of 4.2 kg at birth. Fetus A had a normal developed two upper and two lower limbs with no anomaly of the fingers.

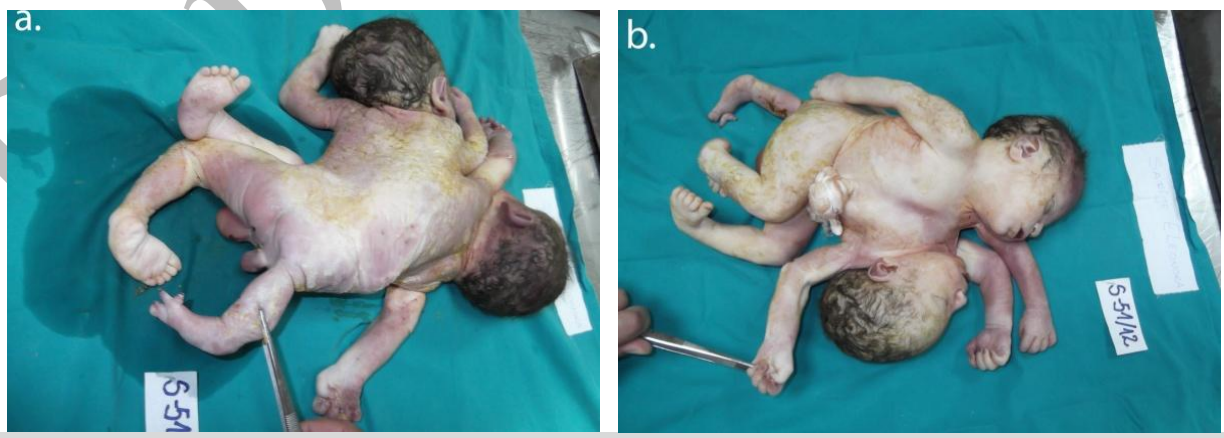

Figure 1. Posterior (a) and anterior (b) view of parapagus twin males.

Both feet were in the position of equinovarus. The upper extremities of fetuses B were normally developed, but there was one bizarre looking rudimentary lower limb, with elongated feet and

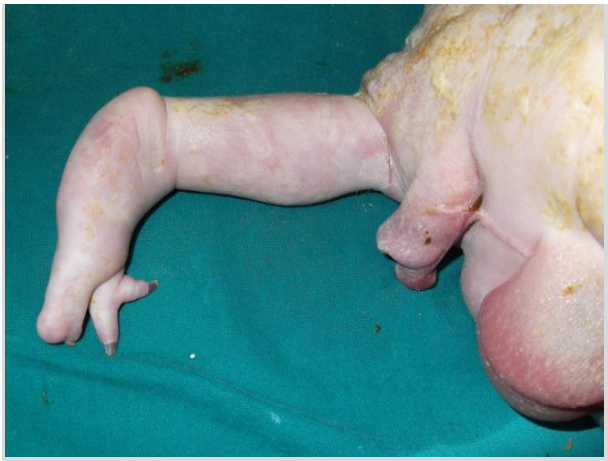

Figure 2. Rudimentary lower limb.

ectodactyly and syndactylic fingers. Three unusual fingers were presented with creations in place of nail plates that had the appearance of claws (Figure 2). Postmortem-made X-ray of 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 the twin A and one testicle in scrotal bag of twin B. There was one anus.

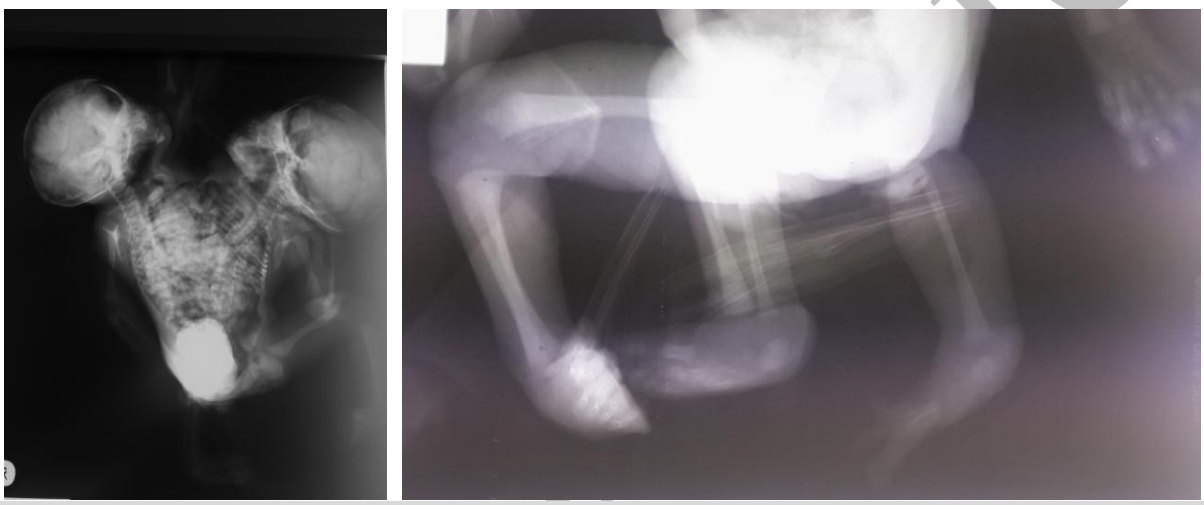

Figure 3. Postmortem X ray shows two separated spines and visible third limb with one bone.

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 the fetus A had a great defect in the diaphragm with the penetration of the liver and intestines into the chest cavity, with severe hypoplasia of both lungs, while the fetus B had normal developed diaphragm. In the thoracic cavity of each fetuses the fully developed heart without any anomaly was 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 the fetus A has a usual morphology of

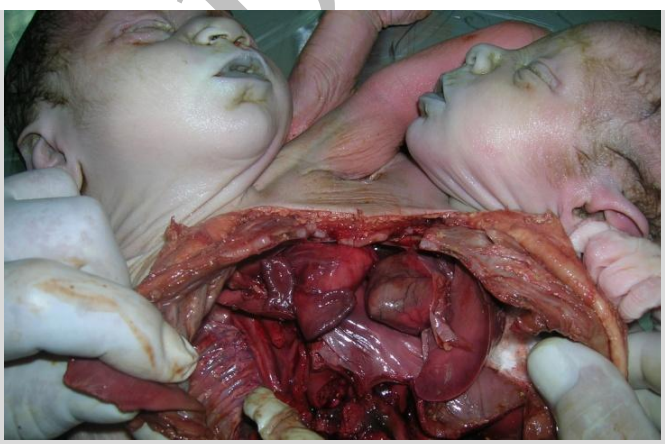

Figure 4. Opened thoracic cavity showing two separated hearts.

thoracic and abdominal aorta and its branches, while the fetal aorta B flowed in the abdominal part of the aorta of the fetus A.

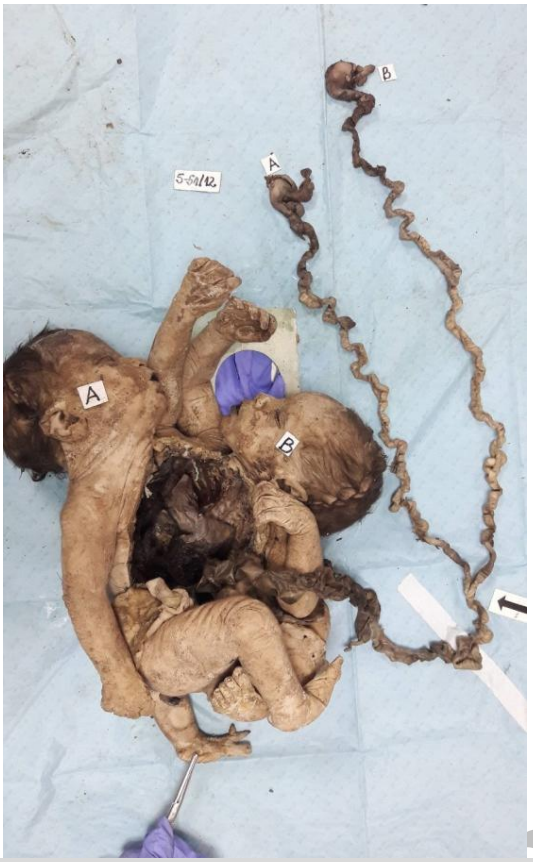

Figure 5. Place of conjunction of two parallel small intestines and separated stomachs.

The abdominal cavity was common to both twins, 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-remaining part of the small intestine and colon, with no anomalies (Figure 5).

Fetal liver were separate in each, where the fetus A liver was placed in the thoracic cavity because of diaphragmal defect. Both of fetuses had a pancreas in appropriate localization. The urinary system was found only in twin A – two apparently normal developed kidney and adrenal gland with two ureters, a urinary bladder and a 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 Chulkhurst from England, 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's Thailand, for which this anomaly is named "Siamese twins" [2, 13, 14].

The most common forms of conjoined twins are thoracopagus and thoraco-omfalopagus, while the incidence parapagus is smaller [7, 15, 16]. It was noted that parapagus more often develops in male twins, compared to other varieties of Siamese twins in general that are more common in female fetuses [2]. The etiology of the disorder is not known yet, but the Siamese twins, with no statistically significant differences can develop in primigravida, or in multigravida, and it is shown that maternal age is not related with this type of twin pregnancy [17]. In our case it was the 20-year-old primigravida, with information of abuse of narcotics and sedatives by both mother and father of twins. The information about the type of narcotics and sedatives, as well as 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 [18], but in our case the question arises whether the abuse of opiates and sedatives had an influence in development of malformated twins.

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

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

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