# Anencephalic conjoined twins with mirror-image cleft lip and palate

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**Summary**

This is a case presentation of a conjoined twin (cephalothoracopagus) pregnancy with anencaphaly and mirror-image cleft lip and palate, affecting the left side of one twin and the right side of the other twin. The pregnancy was terminated at 26 weeks. The case is discussed with information in the literature.

*Key words:* Conjoined twin; Anencephaly; Cleft lip; Cleft palate.

## Introduction

Conjoined twins are a rare complication of monochorionic twinning. The incidence is somewhere between one in 50,000 and 100,000 births (or 1 in 500-600 twin births) [1]. The most famous conjoined twins were Chang and Eng Bunker, born in Siam (now Thailand) in 1811 and joined by a small bridge of union at the umbilicus (omphalopagus). These twins gave rise to the popular term 'Siamese twins' [2]. Classification of conjoined twins is typically based on the fused anatomic region followed by the suffix, 'pagus', to indicate fastened. A simplified classification system for the eight classic types of conjoined twins was proposed by Spencer [3]. Conjoined twinning and cleft lip and palate have been previously reported in the literature [4-7]. Cleft lip and palate in cephalothoracopagus twins has also been reported [8, 9]. Mirror-image cleft lip and palate in thoracopagus twins have been described [10-15].

In this paper, we report a case of cephalothoracopagus conjoined twins with anencephaly and mirror-image cleft lip and palate who were born in Turkey. To our knowledge, this is the first report of such a case in the world.

## Case Report

A 32-year-old woman, gravida 4, para 2, at 26 weeks of gestation was referred to our Perinatology unit for pregnancy termination with a presumptive diagnosis of anencephaly in the fetus. Her past medical history was unremarkable. She did not have any systemic disease, tobacco or alcohol use. Familial history for any kind of anomaly was negative. Obstetric history revealed that she had two healthy children and a term intrauterine unexplained death. She received no prenatal care until the 26$^{th}$ week of gestation. She had no drug usage during pregnancy. The ultrasound examination performed in another hospital revealed anencephalic fetus; thus the patient was referred to our center with a presumptive diagnosis of anencephaly for pregnancy termination. A diagnosis of conjoined twins had been missed by using 2-D sonography. Medical termination of the pregnancy was performed by using misoprostol. A set of conjoined female twins with craniothoracopagus anomaly with a total weight of 1220 grams was evacuated. Anatomical features included development of two faces with mirror-image configuration cleft lip and palate, two brain stems, two vertebral columns, four lower limbs, four upper limbs, two female genitalia, and a single shared anencephalic head (Figure 1, 2). The family refused further investigation, including autopsy, due to their strict religious beliefs.

## Discussion

Conjoined twinning is a rare congenital condition that has an incidence of one in 50,000 births [1]. The embryologic origin of conjoined twinning is still debated. Two contradicting theories exist – the fusion and the fission theories [16]. According to the fission theory, which has been the generally accepted one, during the normal course of monozygotic twinning, division by an unknown stimulus occurs at around 13 days post-fertilization when the embryo is too large to separate fully and remains united at one pole or the other, or at a point between the poles [17]. The fusion theory suggests that the inner cell mass divides fully, but the two monozygotic embryos stay close enough to share the amnion alone or both the amnion and yolk sac. Then, they might come in contact with one another and become reunited (fused) resulting in either ventrally or dorsally conjoined twins [16].

Conjoined twins are typically classified by the point at which their bodies are joined. The most common types of conjoined twins are thoraco-omphalopagus, thoracopagus, omphalopagus and craniopagus. Other less common types of conjoined twins include cephalopagus, synecephalus and cephalothoracopagus. In cephalothoracopagus, bodies are fused at the head and thorax. In this type of twins, there are two faces facing in opposite directions, or sometimes a single face and an enlarged skull. Our case had two faces, two brain stems, two vertebral columns, four lower limbs, four upper limbs, two female genitalia, and a single shared head. Congenital malformations usually occur in conjoined twins. The malformations may or may not be associated with the site(s) of fusion. Most commonly these malformations include

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Fig. 1

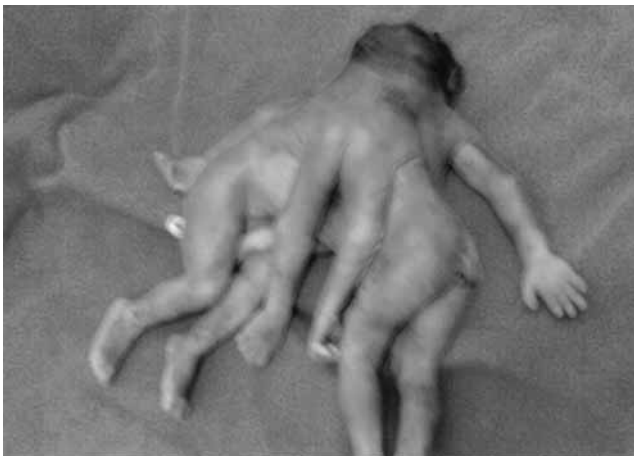

Figure 1. — Conjoined twin with cephalothoracopagus.

Fig. 2

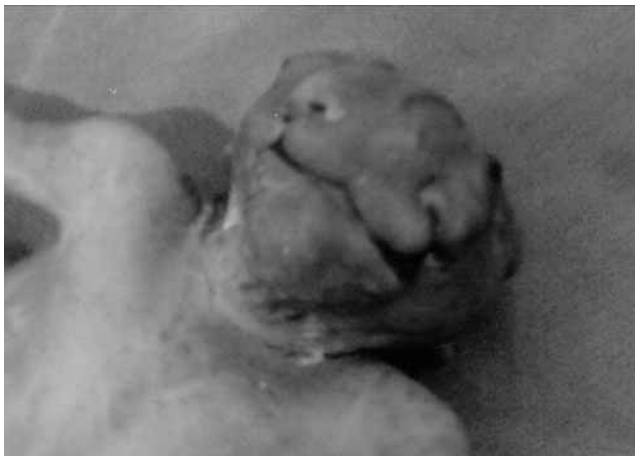

Figure 2. — Conjoined twins showing two faces with mirror-image configuration cleft lip and palate.

neural tube defects and orofacial clefts [18]. Our case had two faces with mirror-image configuration cleft lip and palate and a single shared anencephalic head. Anencephaly and orofacial clefts are multifactorial in origin, arising from both genetic and environmental factors.

The present case as well as other reports on mirror-image clefts in conjoined twins [10-15] supports the fission theory. Mirror-image clefts in conjoined twins have also been suggested to occur from some environmental factors such as poor blood supply [15]. Anencephaly in the case of laterally fused heads was thought to be associated with mechanical difficulty in closing the rostral neuropore [9].

Besides the etiologic factors, diagnosis and route of delivery are important issues in conjoined twins. In our case, the patient was referred with a diagnosis of anencephaly, and the true diagnosis had been missed antenatally. Recently, several case reports have addressed the role of 3-D sonography in the diagnosis [19, 20]. Vaginal delivery was established uneventfully. Multidisciplinary prenatal assessment of conjoined twins is essential to appropriately counsel parents, to manage the pregnancy, and to create an appropriate delivery plan.

To our knowledge, such a case has not been reported to date. We presented this case due to its rarity. This case report supports the importance of fission theory. Further case reports are needed to understand the pathophysiology of this condition.

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