

Conjoined twins with thoracoabdominopagus anomaly in the third trimester: A case report

I ARIKAN, A BARUT, M HARMA, M HARMA, A TANRIVERDI

Citation

I ARIKAN, A BARUT, M HARMA, M HARMA, A TANRIVERDI. *Conjoined twins with thoracoabdominopagus anomaly in the third trimester: A case report*. The Internet Journal of Gynecology and Obstetrics. 2009 Volume 12 Number 2.

Abstract

A patient referred with ruptured membrane was found, by ultrasound and magnetic resonance imaging, to be carrying conjoined thoracoabdominopagus twins. Two live male babies joined at the chest were delivered by caesarean section. The parents refused a separation operation and the twins died from cardiopulmonary arrest after 18 hours. A review of the literature suggests that early diagnosis by a combination of ultrasound and MRI is essential to management, providing prognosis for viability and success of surgical separation and the opportunity for early counselling of parents and termination if indicated.

INTRODUCTION

Conjoined twinning is rare, occurring in about 1% of monozygotic twins, with an estimated incidence ranging from 1:30,000 to 1:200,000 live births (1, 2) and one in 650-900 twin deliveries (3). An increased incidence of 1:14,000 to 1:25,000 is described in various parts of Southeast Asia and Africa (4). Some 40-60% of conjoined twins are reported to be stillborn, and approximately 35% of live births do not survive beyond the first 24 hours (2). There is a reported female predominance in the order of 3:1 (2). Classification is usually made according to the most prominent site of connection: the thorax (thoracopagus; 30-40%), abdomen (omphalopagus; 25-30%), sacrum (pygopagus; 10-20%), pelvis (ischiopagus; 6-20%), skull (craniopagus; 2-16%), face (cephalopagus), or back (rachipagus) (2, 5, 6, 7).

Unexpectedly vaginal delivery of dicephalic parapagus conjoined twins have been reported (8). Here we present our first experience of thoracoabdominopagus conjoined twins diagnosed in the third trimester of pregnancy and briefly review the literature.

CASE REPORT

A 19 year-old gravida 1, para 0, was referred to our department with early membrane rupture at 32 weeks gestation. Her previous medical, surgical, and obstetrical history was unremarkable. There was no family history of twinning on either maternal or paternal sides, and there was no history of medication or X ray. On abdominal

examination, uterofundal height was appropriate to the expected gestational age and corresponded to 32 weeks gestational age. On vaginal examination, external cervical os was 3 cm dilated, and ruptured amniotic membrane was diagnosed. Ultrasonographic evaluation revealed a suspected conjoined twin of the thoracoabdominopagus subtype and decreased amniotic fluid. Doppler measurements on ultrasonography were unremarkable. An MRI was performed in order to confirm the diagnosis (Figure 1). Two live male fetuses joined at the chest, with combined weight of 3.1 kg, were delivered by caesarean section. A single cord had two arteries and one vein, and the placenta was monozygotic monoamniotic. Against all medical advice, the parents refused a separation operation. The babies survived for 18 hours and died due to cardiopulmonary arrest. The parents refused an autopsy examination.

Figure 1

Figure 1. MR image of conjoined twins shows fused thorax.



DISCUSSION

Conjoined twins are mono chorionic mono amniotic twins where twinning is initiated after the embryonic disk and the rudimentary amniotic sac have been formed and fission of the embryonic disc is incomplete (9).

Thoracopagus twins are united face to face from the upper thorax to the umbilicus, with a common sternum, diaphragm, and upper abdominal wall (10). Ninety percent have a common pericardium, 75% have conjoined heart and liver, and a common gastrointestinal tract is found in 50% (11). The treatment options mainly depend on the cardiovascular system anatomy, the success rate of the separation operation being higher where only the pericardium is shared (12). The severity of the cardiac abnormality determines the prognosis, survival, and feasibility of separation. There are no reports of conjoined twins with ventricular conjunction having been successfully separated with both twins surviving (10)

Early diagnosis is the most important factor for management of the pregnancy. With transvaginal ultrasound, a conjoined twin can now be diagnosed at 8 weeks gestational age (13). Diagnostic criteria proposed include: absent separating membrane, conjoined body parts, inseparable bodies or heads despite changes in fetal position, bifid appearance of

the fetal pole in the first trimester, more than three vessels in the umbilical cord, complex structural anomalies, heads or bodies at the same level, hyperextended spine, unusual proximity of the extremities, persistence of the relative positions after movement or at the follow-up scan (7). Polyhydramnios is found in approximately 50–76% of cases, but usually not in the first trimester (9). Recent reports have also presented data on the contribution of 3D ultrasonography to the antenatal diagnosis of conjoined twins (14, 15).

Detailed evaluation of the degree of union and number of shared organs is required to predict the viability and prognosis of the fetuses. Magnetic resonance imaging and computed tomography both provide excellent anatomic and bone detail, demonstrating organ position, shared viscera, and limited vascular anatomy (10), but the consensus appears to be that optimal evaluation is obtained with a combination of ultrasound and magnetic resonance imaging (16, 17). Where the prognosis is poor, early diagnosis enables proper counselling of the family and gives the parents the option of termination.

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Author Information

Ilker ARIKAN, M.D.

Assistant Professor, Department of Obstetrics and Gynecology, Zonguldak Karaelmas University

Aykut BARUT, M.D.

Assistant Professor, Department of Obstetrics and Gynecology, Zonguldak Karaelmas University

Muge HARMA, M.D.

Associate Professor, Department of Obstetrics and Gynecology, Zonguldak Karaelmas University

Mehmet Ibrahim HARMA, M.D.

Associate Professor, Department of Obstetrics and Gynecology, Zonguldak Karaelmas University

Alper TANRIVERDI, M.D.

Associate Professor, Department of Obstetrics and Gynecology, Acibadem Bursa Hastanesi