Acardia acephalus fetus in a twin pregnancy

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The TRAP syndrome or reversed arterial perfusion syndrome is a rare twin-twin transfusion syndrome in monozygotic twins. In this syndrome, the receiving fetus is perfused with reversed blood supply by the pumping fetus. Therefore, the receiving fetus may be affected by malformations such as acardia and acephalus.[1] The severity of the syndrome depends on the kind of vascular anastomosis and its time. Since the pumping fetus is responsible for vascular perfusion of the receiving fetus, therefore, the pumping fetus may be affected by heart failure. There are 2 basic theories regarding acardia acephalus fetuses. During embryogenesis, the normal development of the heart is disturbed due to vascular anastomosis of the twins and reversed arterial perfusion, this in turn, leads to acardia.[2] An abnormal karyotype might also be a cause of acardia.[3] Twin acardia is a rare complication in multiple pregnancies which occurs in 1% of monozygotic twins or approximately one case in 35000 births.[4] In triplet pregnancies, it occurs in approximately one case in 30 pregnancies.[5]

We report a 23-year-old woman, primipara with gestational age of 24 weeks and the chief complaint of labor pain, and premature preterm rupture of membrane for 3 days referred to Amir Hospital of Semnan, Iran. She was married for one year with normal medical background; her husband was her uncle's son (fathers brother's son). On physical examination she had fundal height approximately 26 weeks and 5 cm dilatation, 60% effacement and obvious gush of amniotic fluid from the vagina with unclear presenting part. Uterine contraction was normal with duration approximately 35 seconds and interval approximately 2/5 minutes. Despite normal contraction after 3 hours, no progress was made, and late deceleration occurred. Therefore, cesarean section was carried out. During prenatal care, the ultrasonography had reported that the pregnancy was twin and the first twin was dead and the other alive and healthy. Cesarean section was performed with spinal anesthesia and transverse incision of abdomen. The first fetus was a typical case of acardia acephalus along with polyhydramnios (**Figures 1 & 2**). The second fetus was a normal male premature fetus with oligohydramnios with a normal appearance and Apgar score 4/10 and 500 grams in weight. Despite, immediate resuscitation for the second fetus, the neonate died after 10 minutes. In the first fetus, the lower extremity and pelvis could nearly be distinguished but there was no upper extremity, head, neck, and thoracic organs. The type of anomaly was acardia acephalus twin. The placenta was single, and the fetal membranes were monochorionic. She had no postoperative complications and was discharged after 2 days in good condition.

Twin acardia acephalus is a rare complication,[4] that occurs in monozygotic twins with feto-fetal transfusion syndrome.[1] The predisposing factors of this anomaly are unknown. This twin pregnancy can be divided into 4 groups. 1. Acardia acormus (5%): the fetus only has a head. 2. Acardia anceps (8%): the fetus has some parts of head and face and there are extremities. 3. Acardia amorphous (25%): in this type the fetus is not similar to a human at all, and it has some muscle, bone, cartilage, and other tissues that are covered with skin. 4. Acardia acephalus (62%): fetus has only lower extremities and pelvis and lacks head, upper extremity and chest organs.[3] There are several methods for prenatal diagnosis including: ultrasonography, Doppler velocimetry, and echocardiography of fetus. The hearts can be

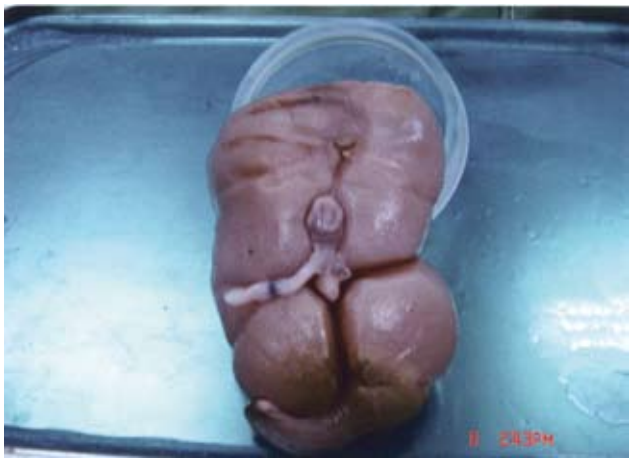

Figure 1 - Anterior view of acardia acephalus fetus.

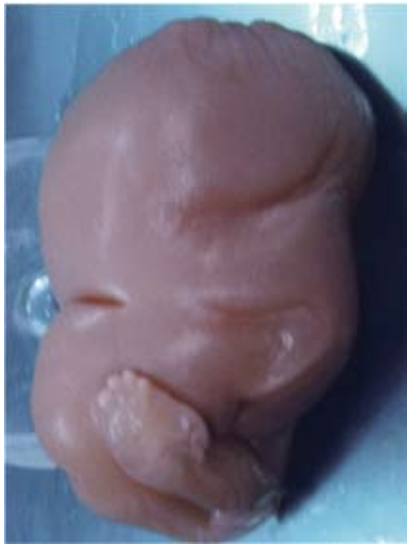

Figure 2 - Posterior view of acardia acephalus fetus.

distinguished with ultrasonography approximately 5-6 weeks into the pregnancy. Diagnosis can be made if there is one heart in one twin and absent in the other. The color Doppler method can show the pulse and twin reversed arterial perfusion of this anomaly can be diagnosed.[5] There are several methods for treatment of this malformation including: 1. Drugs, such as digoxin for the treatment of heart failure in the pumping fetus. 2. Invasive methods based on interruption of vascular perfusion to the receiving fetus to prevent heart failure and other complications in the pumping fetus. These methods consist of: i) embolization and thrombosis of the umbilical cord in the receiving fetus, ii) hysterectomy and delivery of acardia fetus, this is the oldest method,[5] iii) hysterectomy and occlusion of the receiving fetus umbilical cord, iv) occlusion of the umbilical cord by endoscopic methods, v) endoscopic laser coagulation by NDYAG laser.[5] Preterm labor pain, preterm delivery, hydramnios, heart failure during pregnancy, high rates of infections, and hematologic and neurologic complications have been reported in pumping fetuses.[2,3]

In these pregnancies, it will be possible to save the pumping fetus if we diagnose it on time and before delivery.[4] Further monitoring is important in these pregnancies especially when the sonography has reported a dead fetus during pregnancy. Using the color Doppler method showing twin reversed arterial perfusion in acardia fetus, and using endoscopic laser coagulation by NDYAG is the best treatment method in pregnancy,[5] and can be very effective in diagnosis and treatment of these fetuses, it can also prevent multiple complications in the pumping fetus.

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