

Acardiac Twinning With Severe Pulmonary Hypertension And Biventricular Hypertrophic Cardiomyopathy

Akardiak İkiz Eşinde Ciddi Pulmoner Hipertansiyon Ve Biventriküler Hipertrofi

Ayla Günlemez¹, A. Engin Arısoy², Abdulkadir Babaoğlu³, Gülcan Türker¹,
Ayşe S. Gökalp¹

¹Kocaeli University Faculty of Medicine

²Kocaeli University Faculty of Medicine Pediatric Cardiology

³Kocaeli University Faculty of Medicine, Department of Pediatrics

Acardiac twin pregnancy is a rare but serious complication of monochorionic twinning. This acardiac twin gestation complicated by reversible severe pulmonary hypertension and biventricular hypertrophic cardiomyopathy in the pump twin demonstrates the importance of antenatal treatment.

Key Words: *Acardiac twin, Hypertrophic cardiomyopathy, Persistent pulmonary hypertension*

Akardiak ikiz gebelik monokoryonik ikiz gebeliklerin nadir ama ciddi komplikasyonlarından biridir. Burada doğum öncesi izlemi yetersiz olan ve doğumdan sonra ağır pulmoner hipertansiyonla birlikte biventriküler hipertrofik kardiyomyopati tanısı alan bir akardiak ikiz eşi antenatal izlemin önemine dikkat çekmek için sunulmuştur.

Anahtar Kelimeler: *Akardiak ikizlik, Hipertrofik kardiyomyopati, PPHN*

Acardiac twin pregnancy is a rare but serious complication of monochorionic twinning. The acardia phenomenon has an incidence of 1% among monochorionic twins or 1 in 35,000 pregnancies (1). An acardiac twin is a severely malformed fetus that lacks most organs, particularly a heart, but maintains its growth during pregnancy due to its perfusion by the developmentally normal pump twin via a set of arterio-arterial and veno-venous placental anastomoses. The development of a typical pump twin is initially normal; however complications of rapid onset such as plethora, cardiac decompensation, hydrops and intrauterine fetal death usually ensue in the second trimester (2). Without therapy, about 50%-70% of the pump twins die due to congestive heart failure, polyhydramnios and premature delivery (2,3). Two cases of acardiac twin gestation complicated by hypertrophic cardiomy-

opathy have been described in the literature so far (4, 5). We present an acardiac twin gestation complicated by severe reversible pulmonary hypertension and biventricular hypertrophic cardiomyopathy in the pump twin .

Case Presentation

A 22-year-old, primigravida woman was referred to our hospital in her 28th week of gestation because of the ultrasonographic detection of an acardiac acephalic twin and an active pump fetus with normal anatomy and marked polyhydramnios. In the 32nd week of gestation, the mother was found to have premature rupture of membranes and a female infant with fetal distress and weighing 1,750 g, was delivered with cesarean section. Apgar scores of the newborn were 1 and 7 at 1 and 5 minutes,

Received: 25.02.2009 • Accepted: 04.06.2009

Corresponding author

Ayla Günlemez
Cumhuriyet Mahallesi, Sahil Caddesi, Deniz Sokak B Blok D 6
Izmit, 41100 Kocaeli, Turkey
Phone : +90 (262) 226 05 88
Mobile: : +90 (505) 383 68 15
Fax : +90 (262) 303 70 03
E-mail address : aylagunlemez@yahoo.com

respectively. A stillborn acardiac acephalic twin with a weight of 1,400 g was also delivered. Since the infant demonstrated signs of respiratory distress with tachypnea, retractions and cyanosis in the delivery room, she was quickly transferred to the neonatal intensive care unit.

The baby was appropriate for gestational age. There were no dysmorphic features. She had a regular heart rate and rhythm with no murmur. Subcostal and intercostal retractions were noted. Capillary refill time and femoral artery pulse were normal. An initial arterial blood gas analysis revealed severe hypoxia. Chest x-ray revealed hyperinflation of lungs. Whole blood count and serum electrolytes and glucose concentration were within normal ranges. The electrocardiogram showed a QRS axis of 150° with right atrial and right ventricular hypertrophy. The echocardiography revealed severe biventricular hypertrophic cardiomyopathy (Figure 1) with left ventricular dysfunction (fraction shortening: 20%), enlarged right atrium, atrial septum bulging to the left atrium, right to left shunt through a small patent foramen ovale and severe tricuspid regurgitation with a peak velocity of 4.4 m/s, which led to the diagnosis of persistent pulmonary hypertension of the neonate (PPHN). Systolic function is normal and diastolic dysfunction is present.



Figure 1. Parasternal short axis view showing biventricular hypertrophy.

After stabilization, she was placed on a pressure limited, time-cycled ventilator. Over the first three hours of life, the infant needed increasing ventilatory support with up to a FIO_2 of 1.0, a PIP of 22 $\text{cm-H}_2\text{O}$, and a respiratory rate of 60/min. The repeat preductal blood gas analysis revealed a pH of 7.43, PaO_2 of 33 mmHg, PaCO_2 of 24 mmHg, a bicarbonate concentration of 15.9 mmol/L and a base deficit of -5.5 mmol/L. The PaO_2 of preductal blood was 23 mmHg higher than that of postductal blood. The infant also received propranolol (0.1 mg/kg) for afterload reduction and inhaled iloprost (30 ng/kg/dose, every 4 hours) and fentanyl infusion for sedation. Over the next 3 days, she was gradually weaned from assisted ventilation and over the next 12 days she was weaned from nasal prong continuous positive airway pressure. On day 14, iloprost and propranolol were stopped. At the age of 1 year, the echocardiography revealed normal biventricular dimensions and function. The infant is still being followed up with special emphasis on cardiovascular and neurologic systems.

Discussion

It is well established that the pump twin has to circulate more blood volume than in normal conditions in order to perfuse the acardiac twin and that the circulating blood is less oxygenated than normal. Obviously this double volume load constitutes extra work for the surviving heart, which results in dilatation and hypertrophy of the left ventricle and heart failure. Little is known about the course of the normalization process of myocardial hypertrophy, however (2). Szatmari et al (5) described a case with acardiac twin preg-

nancy: the surviving infant presented with short-term heart failure with persistent left ventricular hypertrophy. Chandra et al (4) described another case with biventricular concentric hypertrophy which resolved with propranolol therapy without sequelae by one year of age. It is suggested that biventricular concentric hypertrophy and PPHN was a result of hemodynamic stress and that resolution of the hypertrophy occurred when the stress of the acardiac fetus was removed after birth.

Our case is different in its development of biventricular concentric hypertrophy with severe PPHN, the resolution of the severe pulmonary hypertension in two weeks and the resolution of the biventricular concentric hypertrophy in six months.

PPHN may accompany many types of neonatal chronic intrauterine hypoxia, which may result in hypertrophy of the medial musculature of pulmonary arterioles. Most neonates with PPHN are older than 32 weeks of gestational age and have advanced development of the musculature of pulmonary arterial bed. It has been shown that the degree of the development is directly proportional with gestational age and is appreciable after 32 weeks of gestation (6). Pulmonary hypertension secondary to elevation of left atrial pressure is common in patients with hypertrophic cardiomyopathy; however severe pulmonary hypertension reaching systemic values is unusual in these patients, especially in the absence of severe obstruction and severe mitral regurgitation (7). In the present case, the infant was 32 weeks gestational age and the diagnosis of PPHN was established by echocardiography.

Survival of one of the "pump" fetus

implies the necessity of early diagnosis for the detection of gestational pathology, possible intrauterine interventions and monitoring of the healthy twin. Fetoscopic laser coagulation of placental vascular anastomoses or the umbilical cord of the acardiac twin is an effective treatment of twin reversed arterial perfusion sequence, with a survival rate of 80%, and 67% of pregnancies with surviving pump twins going beyond 36 weeks' gestation without further complications (8). Therapies, includ-

ing conservative treatment and invasive procedures, like intrafetal alcohol chemosclerosis, are directed towards achieving optimal maintenance of pump twins based on clinical presentation (9). Fetal ventricular hypertrophy could not be diagnosed in our patient during routine antenatal care. In the antenatal ultrasonographic examination, diagnosis of the ventricular hypertrophy could be of great help in predicting the fetal and neonatal status.

Our case demonstrates without antenatal treatment acardiac twin gestation complicated by reversible severe pulmonary hypertension and biventricular hypertrophic cardiomyopathy in the pump twin. Effective antenatal treatment could be with surviving pump twins going beyond 36 weeks' gestation without further complications. Therefore the pump twin should be meticulously searched for physical and echocardiographic findings in collaboration with a pediatric cardiologist.

REFERENCES

1. Napolitani FD, Schreiber I. The acardiac monster. A review of the world literature and presentation of 2 cases. *Am J Obstet Gynecol* 1960;80:582-587.
2. Gemert MJ, Umur A, Wingaard JP, VanBavel E, Vandenbussche FP, Nikkels PG. Increasing cardiac output and decreasing oxygenation sequence in pump twins of acardiac twin pregnancies. *Phys Med Biol* 2005;50:33-42.
3. Moore TR, Gale S, Benirschke K. Perinatal outcome of forty nine pregnancies complicated by acardiac twinning. *Am J Obstet Gynecol* 1990;163:907-912.
4. Chandra S, Crane JMG, Young DC, Shah S. Acardiac twin pregnancy with neonatal resolution of pump twin cardiomyopathy. *Obstet Gynecol* 2000;96:820-821.
5. Szatmari A, Anker JN, Gaillard JLJ. An acardiac infant: the extreme form of the twin-twin transfusion syndrome. *Int J Cardiol* 1993;41:237-240.
6. Goldsmith JP, Roca TP. Ventilatory management casebooks. In: Goldsmith JP, Karotkin EH, (eds). *Assisted ventilation of the neonate*. Philadelphia: Elsevier Inc. 2003:507-529.
7. Brilakis ES, Nishimura RA. Severe pulmonary hypertension in a patient with hypertrophic cardiomyopathy: response to alcohol septal ablation. *Heart* 2003;89:790.
8. Hecher K, Lewi L, Gratacos E, Huber A, Ville Y, Deprest J. Twin reversed arterial perfusion: fetoscopic laser coagulation of placental anastomoses or the umbilical cord. *Ultrasound Obstet Gynecol*. 2006;28(5):688-91.
9. Sepulveda W, Corral E, Aiello H, Otano L, Paredes R, Escobar MF et al. Intrafetal alcohol chemosclerosis of acardiac twins: a multicenter experience. *Fetal Diagn Ther* 2004;19:448-452.