Received: 02/02/2022 *Accepted: 20/03/2022*

International Journal of Advances in Biomedical Engineering
Open Access Peer-reviewed Journal
ISSN: 2822-2237
www.ijabe.online
Volume:1, Number:2, Pages:(69-77)
Year:2022

Partial Thoracic Type Ectopia Cordis; a Case Report

***Tugay Tepe*[*1], *Mustafa Özdemir*[1], *Şerif Hamitoğlu*[1], *Ahmet Kurtoğlu*[1], *Hacer Yapıcıoğlu*[1], *Ferda Özlü*[1], *Orhan Kemal Salih*[2], *Eyüphan Gencel*[3], *Ömer Kokaçya*[3], *Anıl Atmış*[4], *Nejat Narlı*[1], *Mehmet Satar*[1]**

**Corresponding Author E-mail:drtugaytepe@yahoo.com*

[1]*Cukurova University, Faculty of Medicine*
Dept. of Pediatrics, Neonatology
01330, Adana, Turkey
[2]*Cukurova University, Faculty of Medicine*
Department of Cardiovascular and Thoracıc Surgery
01330, Adana, Turkey
[3]*Cukurova University, Faculty of Medicine*
Department of Plastic Reconstructive and Aesthetic Surgery
01330, Adana, Turkey
[4]*Cukurova University, Faculty of Medicine*
Dept. of Pediatrics, Pediatric Cardiology
01330, Adana, Turkey

Abstract

Ectopia Cordis (EC) is defined as a congenital defect which babies are born with their hearts outside their bodies. Abnormally located hearth (partially or totally outside of the thorax) mostly causes accompanying congenital anomalies. Although treatment options are limited and mortality rate is relatively high, surgical intervention is one of the most common treatment approaches. This paper presents a case of thoracic ectopia cordis with good outcome and its surgical intervention.

Keywords: Ectopia Cordis; Surgery; Case; Intervention

1. Introduction

Ectopia Cordis (EC) is defined as a congenital defect in which the heart is outside the thoracic cavity. This condition, which can be partially or completely, is called the Pentalogy of Cantrell together with other accompanying anomalies [1]. Based on the location of the heart, it is classified as cervical, cervicothoracic, thoracic, abdominal, and thoracoabdominal [2]. Previous research has established that it is more common in males and has an estimated prevalence of 5 to 8 per million live births [3]. Although the treatment is primarily surgical, the timing varies depending on the accompanying congenital anomalies. This paper discusses the case of thoracic ectopia cordis with good outcome and its surgical intervention.

2. Case Presentation

The case of a male infant, delivered by a normal vaginal route at 37 weeks of gestation was referred to our hospital with the diagnosis of ectopia cordis. The baby was the second living birth from the 4th pregnancy of a 21-year-old mother. It was stated that the mother had 2 abortions of unknown cause and had a healthy 1-year-old son. The parents were first cousins and no prenatal follow-up was performed. The newborn had an Apgar score of 5 and 6 in the 1st and 5th minutes, respectively. The newborn measured 50 cm and weighed 2,900 g, with a head circumference of 35cm. On examination, it was observed that there was no lower part of the sternum in the thorax region, and the apex of the heart was outside (Figure 1).

The patient's thoracic defect was covered with a sterile 3M drape. No additional anomaly was detected in the transthoracic echocardiography of the patient. Thorax tomography revealed a defect of approximately 2 cm in the anterior thoracic wall and a herniated heart apex on the skin from this defect, and it was stated that the sternum was agenetic (Figure 2).

As a result of a joint meeting with the cardiovascular surgery and plastic surgery departments, it was planned to operate the patient gradually at the most appropriate time. The patient underwent repair surgery of the chest wall On the 30th day of hospitalization. It was observed that there were skin adhesions due to epithelialization developing from the periphery where the heart apex was covered with granulation tissue. The granulation tissue on the myocardium without pericardium was scraped. Alloplastic material polytetrafluoroethylene (PTFE) [4][5].5x4 cm defect size was prepared by cardiovascular surgery and sutured to the deep fascia by controlling ventricular pressure. Pectoralis muscle medial was reached by progressing laterally from the border formed by dissection between the pericardium and the surrounding tissue. In the case without sternum and costochondral junctions, dissection was performed over the pectoralis muscle fascia until it reached the anterior axillary area. The perforators from the serratus and lattisimus dorsi muscle to the skin were preserved. After the medialization of the flaps and fixation sutures to the underlying pectoral muscle fascia, subcutaneous and skin suturing

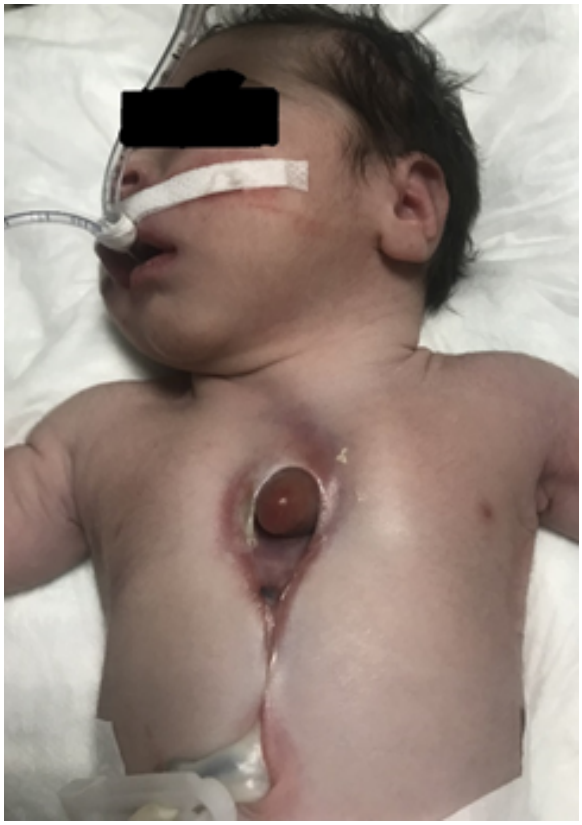

Figure 1: Thoracic type ectopia cordis

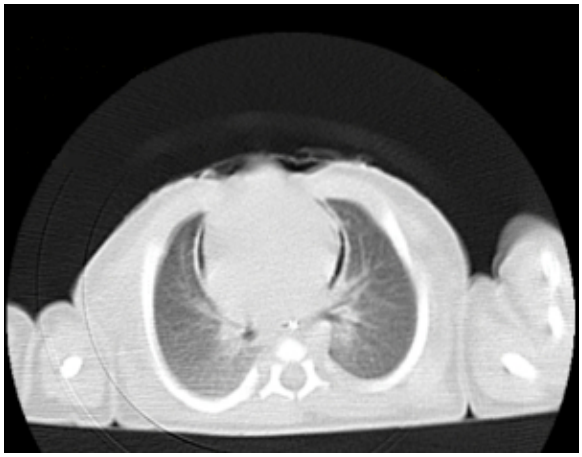

Figure 2: Chest CT findings.

was performed from the sternal notch localization to the umbilicus inferior. In the follow-up of the patient, single z-plasty was performed on the umbilicus for the

midline skin separation due to the compression of the PTFE alloplastic material on the skin. In the first operation for skin tension, the flaps that developed a delay procedure were transformed into bipediculated flaps with incisions made from the anterior axillary line, and the defect area was closed (Figure 3).

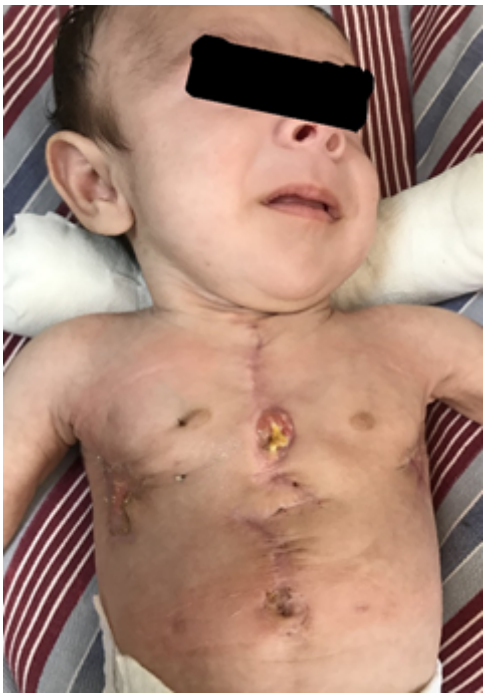

Figure 3: Postoperative view of EC

Thorax reconstruction was planned at a later period. The patient's paradoxical breathing was observed in the postoperative period. Current respiratory distress was attributed to agenesis of the sternum. The patient was discharged on the postnatal 117th day with recommendations. . Informed consent was obtained from the legally authorized representative for publication of this case report and any accompanying images.

3. Discussion

Although EC is rare, it is one of the anomalies that require urgent intervention in the neonatal period since the heart is outside. Its etiology is uncertain, but it is thought that the developmental defects of the sternum and thorax may be related to the embryonic period. Hypotheses regarding the etiology involve midline junction defect and primary descent disorder of the lateral body folds, midline junction defect due to early rupture of the chorion and/or yolk sac, or amniotic band syndrome [4][6]. It may also be associated with some chromosomal disorders such as Trisomy 18 and Turner syndrome [7]. Although chromosome analysis was sent

from the case, it was not evaluated as syndromic based on appearance and physical examination findings. Waiting for the result of chromosome analysis, the array is planned when necessary. EC is classified into 5 groups according to the location of the heart outside the chest. In the cervical type, the heart is located in front of the neck and there is no defect in the sternum. In the thoracocervical type, the heart is located in the cervical region, but there is an opening in the upper part of the sternum. In the thoracic type, the heart is partially or completely outside the thoracic cavity with the absence of the sternum or a wide cleft. In the thoracoabdominal and abdominal type, also called Cantrell's syndrome, the heart emerges from the abdominal cavity through a defect in the diaphragm [8]. The most common type is thoracic and abdominal type, accounting for 90% of known cases [9]. The cervical type is the most severe form and usually results in intrauterine death[10]. According to Smith, it was shown that restricted thoracoabdominal type EC has better results than cases with complete EC [11]. Complications such as heart failure, tamponade, cardiac rupture, endocarditis, and sudden death have been described in patients with EC [10]. However, it can be seen in a single cardiac defect or severe congenital cardiac malformations and affects the prognosis. However, other midline defects such as cranial cleft, cleft palate lip, hypoplastic lungs, diaphragmatic hernias can often accompany [12]. The fact that the heart is outside the thoracic cavity causes paradoxical movements of the lungs, and recurrent infection and hypoxemia are considered the main causes of death due to heart failure [10, 13]. The case had thoracic type EC with sternum agenesis. Only the apex of the heart protruded through the defect. The heart was not covered by the pericardium and was not accompanied by any other midline defect. The case had partial thoracic type EC and the prognosis was evaluated as better. Although paradoxical respiration was observed from time to time, recurrent lung infections did not develop. The absence of congenital heart defect with the partial type defect was evaluated as the reason for the better outcome. Diagnosis can be made very easily with ultrasonography in the prenatal period. If termination is not considered, clinical course and additional anomalies can be determined by MRI and fetal echocardiography [10]. Since the case did not have prenatal follow-ups, the diagnosis was made after birth, and tests were performed in terms of additional anomalies. The treatment of EC is primarily surgical repair. However, the timing of surgery varies according to the type of defect and additional anomalies. A multistage repair is the most common approach for surgical repair depending on the external condition and malposition of the heart. Recently, the multi-step surgical repair strategy is enclosed covering the heart with soft tissue, lowering the heart into the thoracic cavity, repairing additional cardiac defects, if any, reconstruction of the chest wall[14]. Due to the small defect of the case, the defect was first covered with a polytetrafluoroethylene (PTFE) membrane. In the next surgical application, tissue transfer was performed with a skin graft. Thoracic reconstruction was left for a later time.

4. Conclusion

Although EC is rare and the probability of survival is low, accompanying anomalies and the location of the defect affect the prognosis. The reason for the good prognosis in our case was evaluated as the small defect and the absence of accompanying anomalies. Besides, a multidisciplinary approach, including obstetricians, neonatal specialists, pediatric cardiologists, cardiac surgeons, and plastic surgeons, increases the chances of survival.

Rights and permissions

References

[1] J. R. Cantrell, J. A. Haller, M. Ravitch, et al., A syndrome of congenital defects involving the abdominal wall, sternum, diaphragm, pericardium, and heart, Surg Gynecol Obstet 107 (5) (1958) 602–614.

[2] M. R. Harrison, R. A. Filly, P. Stanger, A. A. de Lorimier, Prenatal diagnosis and management of omphalocele and ectopia cordis, Journal of pediatric surgery 17 (1) (1982) 64–66.

[3] N. Pamidi, V. R. Vollala, S. Nayak, S. Bhat, Ectopia cordis and amniotic band syndrome, Archives of Medical Science 4 (2) (2008) 208–211.

[4] T. Humpl, P. Huggan, L. K. Hornberger, B. W. McCrindle, Presentation and outcomes of ectopia cordis., The Canadian Journal of Cardiology 15 (12) (1999) 1353–1357.

[5] A. Dobell, H. Williams, R. Long, Staged repair of ectopia cordis, Journal of pediatric surgery 17 (4) (1982) 353–358.

[6] L. C. Kaplan, R. Matsuoka, E. F. Gilbert, J. M. Opitz, D. M. Kurnit, J. F. Reynolds, Ectopia cordis and cleft sternum: evidence for mechanical teratogenesis following rupture of the chorion or yolk sac, American journal of medical genetics 21 (1) (1985) 187–199.

[7] R. Achiron, M. Shimmel, B. Farber, J. Glaser, Prenatal sonographic diagnosis and perinatal management of ectopia cordis, Ultrasound in Obstetrics and Gynecology: The Official Journal of the International Society of Ultrasound in Obstetrics and Gynecology 1 (6) (1991) 431–434.

[8] R. Kanagasuntheram, J. Verzin, Ectopia cordis in man, Thorax 17 (2) (1962) 159.

[9] S. Puvabanditsin, V. Di Stefano, E. Garrow, R. Wong, J. Eng, J. Balbin, Ectopia cordis, Hong Kong Med J 19 (5) (2013) 447–50.

[10] S. A. Engum, Embryology, sternal clefts, ectopia cordis, and cantrell's pentalogy, in: Seminars in pediatric surgery, Vol. 17, Elsevier, 2008, pp. 154–160.

[11] B. J. Smith, J. N. Flyer, E. M. Edwards, R. F. Soll, J. D. Horbar, S. B. Yeager, Outcomes for ectopia cordis, The Journal of Pediatrics 216 (2020) 67–72.

[12] R. Carmi, R. Parvari, S. Ehrlich, B. Cwikel, Y. Weinstein, Mapping of an x-linked gene for ventral midline defects (the tas gene), Birth defects original article series 30 (1) (1996) 179–187.

[13] A. Hannoun, I. M. Usta, F. Sawaya, A. H. Nassar, First trimester sonographic diagnosis of ectopia cordis: a case report and review of the literature, The Journal of Maternal-Fetal & Neonatal Medicine 24 (6) (2011) 867–869.

[14] P. Sadłecki, M. Krekora, G. Krasomski, M. Walentowicz-Sadłecka, M. Grabiec, J. Moll, M. Respondek-Liberska, Prenatally evolving ectopia cordis with successful surgical treatment, Fetal diagnosis and therapy 30 (1) (2011) 70–72.