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CASE REPORT

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Cardiosurgical aspects of the ectopia cordis

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Abstract

The case is presented of a 2710 g newborn female with prenatally diagnosed ectopia cordis. Postnatal transthoracic echocardiography showed the presence of a ductus-dependent intracardiac lesion. The neonate was treated surgically on the first day of life. Different surgical aspects of ectopia cordis are discussed. (Folia Cardiol. 2006; 13: 436–438)

Key words: ectopia cordis, neonates, surgery

Introduction

Ectopia cordis (EC) is a very rare congenital heart anomaly (1 per 5.5–7.9 million births). Untreated it soon leads to death. Ectopia cordis can be isolated or may coexist with other extracardiac and intracardiac lesions (ventricular septal defect, atrial septal defect and lesions of the abdominal wall) [1–3]. There are also numerous chromosomal aberrations which coexist with EC [4].

There is partial or total EC when there are only the great arteries in the chest [3]. It is a rare condition and there are a variety of surgical methods for treating it, involving a single or multi-stage approach. The surgical technique depends on the type of EC and the coexistence or otherwise of heart defects or defects in other organs. The prognosis in EC is poor and there are only a few reports in which a patient with EC survives to long-term follow-up after surgery [1–5].

Case report

Between October 1997 and October 2004 one neonate with EC was operated on in the Department of Pediatric Cardiac Surgery of the University of Medical Sciences, Poznań.

The patient was a one-day-old female with a body weight of 2710 g; she was the third pregnancy (third delivery) and had been born by caesarean section because of a heart defect (EC) in the Department of Gynaecology and Obstetrics of the University of Medical Sciences, Poznań. Ectopia cordis had been diagnosed at 14 weeks of pregnancy during gynaecological ultrasound examination. The neonate had an Agpar score of 1 and 5 points in the first and fifth minute of life. The patient was intubated with a 3.5 mm tube because of respiratory insufficiency in the first minute of life and was mechanically ventilated. A bandage with a warm solution of 0.9% NaCl was put on the heart, which was outside the chest.

The neonate was transferred from the Department of Neonatology to the Department of Pediatric Cardiac Surgery in Poznań. The patient's general status was stable. In laboratory examination pO_2 and pCO_2 in the arterial blood was about 40 mm Hg and saturation equalled 80%; urine output was normal. After admission the patient was put in a heated incubator and treated with 5% albumin, 5% glucose and antibiotic intravenously administrated. The neonate was also treated with Prostin (0.02 μg/kg/min). The stomach was intubated with a nasogastric tube to decontaminate the digestive tract. Echocardiography, which was performed with antiseptic restrictions, showed additional, coexisting heart defects: pulmonary atresia (PA), ventricular septal defect (VSD), and patent ductus arteriosus (PDA).

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Echocardiography was performed in difficult technical conditions because each touching of the heart caused patient instability. The ultrasound examination of the head showed a normal image of the brain. There were no symptoms of bleeding.

When the necessary laboratory tests had been performed and blood from the donor prepared, the girl was transported to the operating theatre.

Surgical technique

After the surgical field had been prepared in the normal way, almost total schisis of the sternum (instead of the manubrium sterni) was recognised, with the heart outside the chest. A longitudinal cut was made of the skin and then of the sternum. The pericardium was separated from the musculocutaneous flaps and also from the costal arch. The pleura was opened and the heart moved into the chest. Drainage was inserted under the sternum. All the stitches were inserted at the edges of the sternum.

Additionally the musculocutaneous flaps were dissected bilaterally from the ribs and stitches were also placed on the muscles. At this time aspiration drainage was opened and the sternotomy closed. No haemodynamic disturbances were observed.

Continuous intravenous infusion of dopamine (9.8 μg/kg/min) and dobutamine (19.7 μg/kg/min) was started during surgery to keep the systemic pressure above 50 mm Hg. Echocardiography was performed in the operating theatre. This revealed a small narrow left ventricle with normal contractility. A patent ductus arteriosus was re-confirmed.

The patient was transferred to the Pediatric Intensive Care Unit in a stable condition.

Discussion

Ultrasound diagnosis of EC can easily be achieved in the first trimester of gestation as was confirmed by our case [4, 6].

A uniform algorithm of treatment remains to be defined according to Alphonso et al. [5], which is why various surgical methods are used and the choice depends on each individual case. The outcomes of treatment are a function of the degree of ectopy and accompanying lesions. The thoracic type of ectopy and a cephalad orientation of the apex with the presence of extracardiac anomalies is predictive of an unfavourable outcome. Patients with isolated EC are more likely to survive [6]. It should be noted that compression over the heart and hypoplastic lungs must be avoided during reconstruction of the thoracic chest wall [1–6].

Operating approaches may be one-stage [1–3] or multi-stage [1–5]. A one-stage correction by Amato et al. [3] included: 1) the placement of the heart in the thoracic cavity, 2) sternum and thoracic chest wall reconstruction, 3) covering of the "naked heart". Indispensable prerequisites of heart repositioning are haemodynamic stability and the absence of any signs of metabolic acidosis in the neonate. A leading apical stitch attached to the diaphragm can be helpful in repositioning the heart and in its stabilisation in the thoracic cavity [2, 3, 5].

Augmentation of the thoracic cavity is necessary [3] in repositioning of the heart, especially where there are coexisting defects of the diaphragm and/or abdomen wall. This situation is associated with the need for phrenic nerve division leading to diaphragm palsy, which results in prolonged mechanical ventilation and a second surgical procedure, that of diaphragm plication.

A two-stage operation includes: I. skin closure of the thorax defect and II. reconstruction of the layers of the chest wall after stabilisation of the patient and augmentation of the thoracic cavity. Morales et al. [1] recommends moving on to the second stage in the second year of a child's life. In the event of a shortage of skin to cover the heart the use of a PTFE (politetrafluoroethylene) [1, 3, 5] membrane or pulmonary homograft is recommended [4].

The sternal schisis is a common element of EC. According Daum and Zachariou [7], it can be total or partial (in the lower or the upper part). Reconstruction of total sternoschisis is as follows: after skin incision and separation of the sternal margins the perichondrium is incised longitudinally and dissected free. Single stitches are put over the free margins, beginning from the lower part of the sternum. Abdominal muscles are adopted with the use if absorbable sutures. After placement of all the stitches they are gradually pulled out and tied in order to create safe conditions for circulatory and respiratory adaptation. At the same time the patient is strictly monitored.

Daum converts partial into total sternoschisis by division of the adherence site. The separated sternal edges are then approached by PDS sutures. Burton [8] fills bone defect with autologous graft from the costal angle, Asp and Sulamaa [9] with bone harvested from the patient's skull.

Authors have underlined the fact that neonatal corrections do not usually need following corrective chondrotomy owing to the considerable elasticity of the ribs and clavicles.

In the case of the coexistence of intracardiac defects the treatment modality depends on the

experience of the medical centre and the results of the management of selected malformations in neonates. A multi-stage strategy of treatment was applied in the case presented. During the first stage the heart was repositioned into the thorax and the surface defect closed. Application of a Blalock-Taussig shunt was postponed because of the high risk of infection [5], the PDA that was confirmed in the ECHO examination, and the need for sufficient arterial blood oxygenation (pO_2 = 40 mm Hg, SpO_2 = 80%).

Morales et al. [1] recommend pulmonary artery banding in the case of coexistent VSD with symptoms of circulatory insufficiency and a Blalock-Taussig shunt in the case of symptomatic Fallot's tetralogy.

The literature reveals only one case of single-stage simultaneous radical correction of EC and intracardiac defect (DORV, double outflow right ventricle) in a neonate. This child died from a generalised infection [5].

Tokunaga et al. [10] successfully performed one-stage neonatal treatment of EC accompanied by single ventricle (DORV with hypoplastic left ventricle and pulmonary stenosis), applying a 4 mm Blalock-Taussig shunt. At the age of 2.8 years the child underwent the second operation, the bi-directional Glenn and pulmonary artery plasty followed by a successful extracardiac Fontan operation at the age of 4 years. Hornberger et al. [11] presented two cases of children with EC and single-ventricle physiology treated with the Fontan method. Both authors recommend the staged treatment strategy.

Other authors suggest heart transplantation as the method of choice in case of EC coexisting with serious intracardiac anomalies.

References

1. Morales JM, Patel SG, Duff JA, Villareal RL, Simpson JW. Ectopia cordis and other midline defects. Ann Thorac Surg, 2000; 70: 111–114.
2. Samir K, Ghez O, Metras D, Kreitmann B. Ectopia cordis, a successful single stage thoracoabdominal repair. Interactive Cardiovasc Thorac Surg 2003; 2: 611–613.
3. Amato JJ, Zelen J, Talkwalkar G. Single-stage repair of thoracic ectopia cordis. Ann Thorac Surg, 1995; 59: 518–520.
4. Sharma VK, Kiran U, Sharma J, Kapoor PM, Saxena N. Challenges in the management of ectopia cordis. J Cardiothorac Vasc Anaesth 2001; 15: 618–623.
5. Alphonso N, Venugopal PS, Deshpande R, Anderson D. Complete thoracic ectopia cordis. Eur J Cardiothorac Surg, 2003; 23: 426–428.
6. Respondek-Liberska M, Janiak K, Włoch A. Fetal echocardiography in ectopia cordis. Pediatr Cardiol, 2000; 21: 249–252.
7. Daum R, Zachariou Z. Total superior sternal clefts in newborns: a simple technique for surgical correction. J Ped Surg, 1999; 34: 408–411.
8. Burton JF. Method of correction of ectopia cordis. Arch Surg, 1947; 54: 79–81.
9. Asp K, Sulamaa M. Ectopia cordis. Acta Chir Scand, 1959; 118: 392.
10. Tokunaga S, Kado H, Imoto Y, Shiokawa Y, Yasui H. Successful staged-Fontan operation in a patient with ectopia cordis. Ann Thorac Surg, 2001; 71: 715–717.
11. Hornberger LK, Colan SD, Lock JE, Wessel DE, Mayer JE. Outcome of patients with ectopia cordis and significant intracardiac defects. Circulation, 1996; 94 (Suppl II): II-32–II-37.