

IMAGES

in PAEDIATRIC CARDIOLOGY

Chelo D,^{1,2} Mbassi Awa HD,^{1,2} Nguéfack Dongmo F,^{1,2} Menanga AP,³ Ngo Um S,^{1,2} Nde Kamgne M,⁴ Koki Ndombo PO.^{1,2} Ectopia cordis. A report of two cases in Cameroon. *Images Paediatr Cardiol* 2014;16(4):1-6

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Abstract

We report two cases of ectopia cordis in two children aged one day and twenty months respectively. A one day old newborn had complete thoracic ectopia cordis associated with an internal cardiac defect and severe thoracic and abdominal wall malformations. Our centre does not have the facilities to manage complex congenital defects and prior to being transferred to a cardiac centre, the neonate died on the second day of admission. A 20-month old baby had partial ectopia of the heart and a defect in the abdominal wall. He had no major congenital cardiac defect and has remained clinically stable with no life threatening symptoms.

MeSH: ectopia cordis; congenital; respiratory distress; cameroon

Introduction

Ectopia cordis is a congenital disorder characterized by the aberrant position of the heart, outside its normal mediastinal location. It is due to a defect in heart migration during embryogenesis, with the heart being found partially or completely out of the rib cage.

This is an extremely rare malformation in the human race, of very poor prognosis with an incidence of 5.5 to 7.9 per million live births.¹⁻²

Depending on the final location of the heart, 4 types are classically described: the thoracic form (65%); thoraco-abdominal (20%); abdominal (10%); and cervical (5%).³

Case reports

Case 1

E.M. a male newborn, weighing 3150 grams and measuring 50 cm was seen at the 13th hour of life at our hospital in Yaounde. He was a product of a non-consanguineous union, delivered by caesarean section at 41 weeks' gestation on account of placenta praevia at a clinic in the town of Buea, in the south-west region of Cameroon, about 400 km from Yaounde. He was referred to our Centre on account of an extrathoracic heart, associated with an anterior defect of the chest wall and abdomen.

Mother, 23 year old primigravida, had received antenatal care throughout the pregnancy. Screening for HIV, syphilis, HBV, rubella and toxoplasmosis were all negative at the 2nd month of gestation. A fetal ultrasound done at third month of pregnancy reported no abnormality. She had quarterly malaria prophylaxis with sulfadoxine + pyrimethamine at the recommended doses as well as iron and folic acid. She did not take any other medication in pregnancy. The mother does not smoke or drink alcohol.

Father is a 24 year old mason. There was no family history of congenital heart disease.

Examination of the newborn upon arrival revealed chest wall abnormalities. His heart was externalized, covered only by the pericardium (Figure 1), the apex oriented towards the tip of the left shoulder and was beating regularly at 156 beats per minute. The sternum was not visualized and he had severe sternal retraction on inspiration. There was no skin covering down to the umbilical region. A peduncle of about 5cm in diameter and 7cm long connected the heart to the chest. He was in severe respiratory distress with a Silverman score of 8/10. There was no cyanosis, and oxygen saturation taken from the finger was 90%. There were no dysmorphic features or other clinically obvious abnormalities.

Figure 1 : Case 1, new born baby with the heart completely exteriorized, covered uniquely by the pericardium



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An echocardiogram showed a 10mm perimembranous ventricular septal defect. Cardiac chambers were however well developed and myocardial contraction was fairly good. The immediate management of the patient consisted of oxygen therapy by face mask at a rate of 3 liters/minute. Aseptic measures were observed. The heart was covered with sterile gauze that was regularly soaked with normal saline.

A nasogastric tube was inserted and enteral feeding was not commenced. The patient received an intravenous infusion of 10% dextrose solution to which electrolytes were added.

In addition, antibiotic prophylaxis with ceftriaxone and gentamycin at a dose of 50 mg/kg and 5 mg/kg respectively per day were commenced. The only cardiac surgical center close to our institution is located at about 600 km from Yaounde. The surgeon had been contacted by telephone for a possible transfer but the baby died 24 hours after admission of severe respiratory failure.

Case 2

M.A. is a 20 months old male infant who presented with a pulsatile swelling in the anterior abdominal wall. He is the only child of a 29-year old housewife and a 34-year old man who is a primary school teacher in a non consanguineous marriage. They drink alcohol occasionally but do not smoke.

M.A. was born at term at a clinic in the outskirts of Yaounde with a birth weight of 3,000 grams. The health personnel who took care of the newborn noted the presence of a pulsatile mass on the anterior surface of the abdomen which was covered with a hyperpigmented skin. The mass was thought to be an umbilical hernia requiring no active intervention.

A family friend advised the parents to consult our services. On physical examination, his anthropometric measurements were normal for his age (weight 12.5kg, height 85 cm, head circumference 48 cm). We noted a pear-shaped midline abdominal mass extending from the epigastric region to the umbilical region. It measured 20 cm long and 12 cm in diameter (fig 2). The mass was completely covered by hyperpigmented skin.

Figure 2 : case 2, 20 month old infant partial ectopia cordis, covered by the skin. Voluminous abdominal mass



In the epigastric hollow, there was an oblong mass measuring 5 x 7 cm, extending into the thorax, firm to palpation, pulsating at the rhythm of the patient's radial pulse at a rate of 105 beats per minute. The rest of the mass had a soft consistency. On auscultation, bowel sounds were heard. The heart sounds were heard over the precordium and they were normal. Examination of the lungs revealed no abnormalities. There were no dysmorphic features or other abnormalities noted.

A chest X-ray has shown a slightly enlarged heart. An abdominal ultrasound revealed no intraabdominal organ abnormality. There was a herniation into the abdominal cavity through the diaphragm, containing a pulsatile mass representing a portion of the left ventricle.

Echocardiography showed an alteration in the cardiac morphology. The left ventricle appeared thin and elongated, measuring 41.1 mm x 15.2 mm compared to the right ventricle, 44.6 mm x 24.3 mm. No other structural abnormality of the heart was noted and cardiac function was satisfactory. The child was transferred for surgery to a European hospital.

Discussion

Ectopia cordis is one of the extremely rare congenital malformations. It accounts for less than 0.1% of all congenital heart defects.⁴ Our center receives at least thirty thousand children per year with various pathologies. A register of congenital heart diseases was established in January 2006 (it includes 1434 patients to date) and the 2 presented patients are the first cases of cardiac ectopia recorded in our Hospital.

The first case published in Cameroon is that described by Tantchou et al at Cardiac Surgery Center of Shisong in 2009.³ A few isolated cases have been reported in the literature.

Heart ectopia is often associated with other abnormalities involving both the heart and other organs. The set can form a well-described syndrome called pentalogy of Cantrell, which has a poor prognosis.⁵⁻⁶ This syndrome in its complete form includes an omphalocele, anterior diaphragmatic hernia, ectopia cordis, a sternal split, and cardiac defects (VSD, ventricular diverticulum, tetralogy of Fallot).

The first patient we described had ectopia cordis, a complete absence of the sternum, a ventricular septal defect, a defect in the abdominal wall above the umbilicus characterized by absence of a cutaneous covering. These four anomalies correspond to a pentalogy of Cantrell Class 2.⁷

The natural history of cardiac ectopy in its severe form is a progression to death within few hours or days.^{1,4}

Our first patient died after 37 hours of life. Kasole et al⁸ in a case described in Congo reported a patient with a lifespan of 9 hours. However, Tantchou presented a case where the diagnosis of ectopia cordis was made at the age of 7 months. The latter author found a heart in the thoraco-abdominal position, completely covered by skin. Furthermore, there were no other heart abnormalities found as seen in our second reported case. In addition, their patient has been treated in a Western specialist center where surgery was performed under optimum conditions.

Our first patient was born in a peripheral health center with a completely exteriorized heart. He was by public transport and travelled over a very long distance with no attention paid to asepsis. The risk of infection was therefore high. In addition, a total absence of his sternum would lead to instability of the chest wall therefore causing severe respiratory distress. This context could leave very little chance of survival in the absence of rigorous resuscitation and respiratory support.

Our second patient had a heart in the thoraco-abdominal position as that described by Tantchou et al. It was completely covered by skin and apart from the dysmorphia of the left ventricle, no other cardiac anomalies were notable. He leads a normal full life as evidenced by height and weight growth that are adequate for his age, and he is awaiting surgery.

Conclusion

We described two cases of ectopia cordis, one with a very poor prognosis, involving severe intracardiac and extracardiac defects who lived less than 48 hours, and a second case with a better prognosis having no anomaly associated with the defect. He had normal growth parameters at 20 months of life. Facilities in our centre are not adequate for the management of the neonate.

Beyond its rare nature, this anomaly requires expert management, which requires a multidisciplinary approach at all stages. We believe that the second patient will benefit from intervention in a more appropriate environment.

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