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Case report

Ectopia cordis: A case report of pre-surgical care in resource-limited setting

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ABSTRACT

Introduction and importance: Ectopia cordis is a rare congenital malformation of thoracic midline fusion that presents as location of the heart outside the open chest cavity. This presents as a surgical emergency and demands early and specialized intervention. Particularly in resource-limited settings, where prenatal ultrasonography screening is not done, these children are often born in facilities without the capability of managing such conditions definitively, necessitating them to be referred to a specialized centre. At lower health facilities, the challenge is in ensuring that the child is kept stable and protected from infection until they can reach a centre with the facilities required for care. This report describes the management give to such a child until they were successfully handed over to a cardiac institute.

Case presentation: We present a newborn male baby delivered at term to a mother from a low socio-economic background with his heart and abdominal viscera outside the thoracic and abdominal cavity. Despite presenting at a centre without cardiac surgery facilities or cardiologists, they were sustained until referral.

Clinical discussion: Ectopia cordis is a rare congenital anomaly characterized by defect in the fusion of the anterior chest wall resulting in the abnormal extra-thoracic location of the heart. Five types exist; cervical type with worst prognosis, attempts can be made to re-locate the heart and close the thoracic defect surgically.

Conclusion: Even with limited resources, it is possible to provide the basic care necessary to sustain a child with this complex anomaly until definitive management can be provided.

1. Background

Ectopia cordis (EC) is a rare congenital malformation of the anterior chest wall resulting in an abnormal extra-thoracic location (partial or complete) of the heart [1–3]. Advances in surgical techniques have enabled these malformations to be corrected in a number of ways. We present a newborn baby with complete EC in a resource-limited setting and how this child was sustained until they could be moved to a better-resourced centre.

This work has been reported in line with the SCARE 2020 criteria [7].

2. Case presentation

This eleven-hour-old male baby was referred because of location of the heart and abdominal organs outside of the body. There was no bleeding reported, and the baby was said to breastfeed well. The baby was delivered at home vaginally at 37 weeks gestation age by dates after spontaneous rupture of membranes and cried immediately.

The mother booked into antenatal clinic at six months of gestation, attended three times, was normotensive, not diabetic, and HIV negative. She was routinely supplemented with folate and iron. She was 21 years of age, and this was her second child, her firstborn being three years old. The mother denied family history of any congenital anomalies.

Upon examination, the baby was awake, pale, not jaundiced, not cyanosed and not dyspneic. The axillary temperature was 35.4 °C, respiratory rate was 37 breaths per minute, heart rate of 144 beats per minute, and saturating at 96% on 2 L/min of oxygen.

The baby's chest and cavity and upper abdomen were open, exposing a beating heart and abdominal viscera, covered by a thin membrane. The baby had a webbed neck on the right side and had normal male genitalia (Fig. 1).

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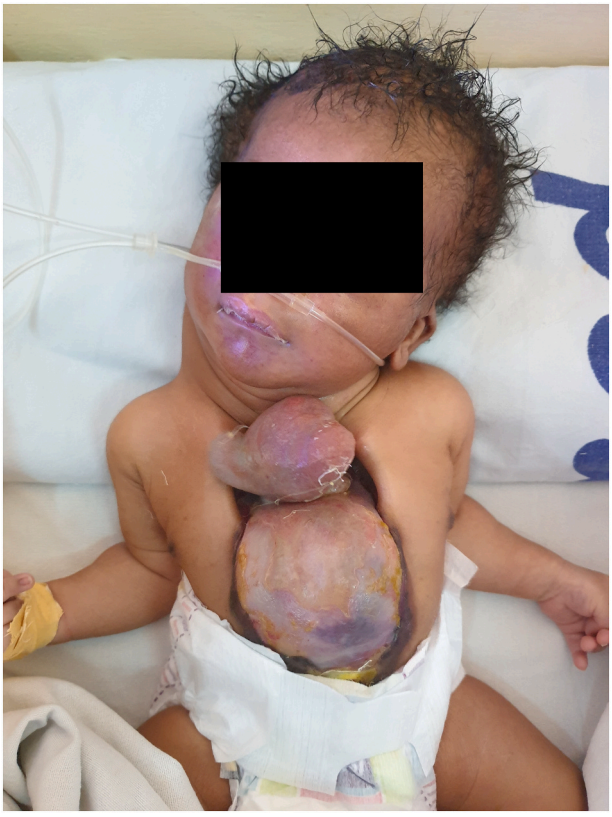

Fig. 1. Photograph showing type-4 Ectopia cordis and webbed neck more towards the right side.

The baby had normal vesicular breath sounds and normal primitive reflexes. The working diagnosis was Ectopia cordis with omphalocele.

A blood count revealed a leucocyte count of 10.35 $\times$ 10^9/L, with elevated monocytes of 1.42 $\times$ 10^9/L, a haemoglobin level of 16 g/dL and a low platelet count of 61 $\times$ 10^9/L. Blood culture was negative.

We kept the baby in an incubator in the neonatal unit to maintain temperature and kept on oxygen. The omphalocele was covered with a sterile moist dressing to prevent desiccation. The child was also kept onantibiotics due to the high risk of infection. The child was fed expressed breast milk via a nasogastric tube.

Preparations were made, and the patient was transferred to Jakaya Kikwete Cardiac Institute (JKCI) on day 4. Unfortunately, after five days at JKCI, the baby had a cardiac arrest, and resuscitation was unsuccessful. Surgery to correct the defect and relocate the heart had not yet been done.

3. Discussion

EC is a rare congenital malformation that was first observed 5000 years ago [2]. It is characterized by a defect in the fusion of the anterior chest wall resulting in the extra-thoracic location of the heart. EC's prevalence is estimated at 5.5–7.9 per million live births with a higher frequency in females [1,2]. Cantrell's pentalogy (a deficiency of the anterior diaphragm, a midline supraumbilical abdominal wall defect, a defect in the diaphragmatic pericardium, congenital intracardiac anomalies and a defect in the lower sternum) has been linked with EC [1]. It is thus worth assessing for other anomalies when EC is identified (Table 1).

Our child had type-four. His sternum was bifid, his heart extra-thoracic, and his anterior abdominal wallwas defective.

The cause of EC is unknown. However, EC is associated with other intracardiac anomalies such as ventricular septal defect, atrial septal defect and tetralogy of Fallot. Due to resource limitations, we were not able to screen for intracardiac lesions prior to referral. EC is also linked with non-cardiac anomalies such as cleft lip and palate, pulmonary hypoplasia, neural tube defects and chromosomal abnormalities like Trisomy 18 and Turner syndrome [1,2,4]. Our patient was suspicious of Noonan syndrome due to the webbed neck, but we could not do genetic analysis to confirm this.

Table 1
Classification of EC [1,3].

Type	Description	Incidence (%)
1. Cervical	Heart is located in the neck with intact sternum	5
2. Thoracocervical	Heart is partially in cervical region,and upper sternum is split	65
3. Thoracic	Sternum is wholly or partially split, and heart is partially or completely outside	
4. Thoraco-abdominal	Usually accompanies Cantrell's syndrome (distal sternal defect, midline supraumbilical defect, ventral diaphragmatic hernia,defect in the pericardium and communication into the peritoneal cavity, and congenital intracardiac defect)	20
5. Abdominal	Heart passes through a diaphragmatic defect to enter the abdominal cavity	10

Diagnosis of EC can be made by an obstetric ultrasound as early as 10–12 weeks [3]. 3D ultrasound and Doppler aid to understand the complex anatomy [4]. Without ultrasound screening during pregnancy, EC is missed till birth, as was the case in our child. Once identified, magnetic resonance imaging allows for better visualization of defects and planning of management [4]. Resource constraints however prevented this from being availed to our patient, whereby this could help to detect early and plan for delivery at centre with adequate resources for care.

Prognosis largely depends on the type of EC and the associated malformations. Cervical type is said to be incompatible with life. Thoracic EC has poor prognosis compared to other non-cervical types, but some survive [5]. Many succumb to the malformations with in the first few hours of life [1]. That our patient managed to survive for one day before presenting to hospital and four days before transfer, speaks towards the potential for good prognosis.

Surgery aims to close the defect and cover the exposed viscera by primary chest wall closure or bone/cartilage grafts or prostheses such as silastic, marlex mesh or acrylic plaques. Intracardiac defects are corrected before closure [2,3].

Despite the reported high mortality rates, successful corrective or palliative operations can be performed during neonatal, infancy or childhood periods [6]. Two-staged repair allows tissue coverage and allows hemodynamic stability of the patient in terms of cardiac physiology and toleration [6]. Thoracic type EC presents a surgical challenge, and the reported survival after birth is 36 h on average due to intracardiac defects [6]. Delayed repair is advisable in certain cases to allow growth of the thoracic cavity, and relocation of the heart is accomplished by two years of age [6].

4. Conclusion

Though the prognosis is generally poor, successes are growing with surgical advances. Prompt recognition and intervention to prevent complications such as hypothermia and infection are important at all levels to allow the patient to carry safely to surgery, whether early or delayed.

Consent

Written informed consent was obtained from the child's mother for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this

journal on request.

Ethical approval

Ethical approval obtained from the Departments of General Surgery and Pediatric, Kilimanjaro Christian Medical Centre.

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Authors' contributions

JL, HC, and RP conceptualizedand drafted the manuscript. KC, DM and RM reviewed the patient medical records. All authors have proofread and approved the final manuscript.

Guarantor

JL accepts full responsibility for the work and the conduct of the study, had access to the data, and controlled the decision to publish.

Research registration

N/A.

Declaration of competing interest

The authors declare they have no conflicts of interest.

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