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INJURIES OF EMBRYONIC LAYERS- CONGENITAL MALFORMATIONS

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Abstract

Ectopia Cordis is a rare malformation in which the heart develops outside the chest. This is sometimes covered by the skin, sometimes only by the pericardium membranes. The causes of this malformation are supposed to be of genetic origin, defective, infectious or other. The myocardium develops from mesoderm, so a lesion at this level could be the cause of this malformation. Necks in the uterine cavity could be the cause of this lack of cord entry into the chest cavity. Theories about this malformation involve rupture in the early embryogenesis of the corion and the Yolk sac. The main cause is the damage of the mesoderm with the lack of sternum closure and implicitly ectopic position of the heart. This malformation is treated in specialized centers for pediatric surgery, but their rarity does not allow very rapid development of surgical techniques, experimental methods are still widely applied and depend on the surgeon's inventiveness and training.

Key words: ectopiacordis, congenital malformation, embryogenesis

INTRODUCTION

The exact etiology remains unknown, but the lateral trunk anomalies seem to be involved(Amato J et al, 2000). Normally, the side wall of the body is responsible for fusion on the midline of the ventral wall (Sadler TW ,2010). Ectopia Cordis is also associated with other malformations such as septal

atrial defect, ventricular septal defect, Fallot tetralogy, atrial tricuspidia, double outlet right ventricle (*Bernstein, D ,2011)*. Its frequency is estimated to be between 5.5-7.9 per million live births (Shad, J, 2012). Treatment of these cases involves care in the 35 degree C incubator at a suitable position, amongst maneuvers is the coverage of the heart and the omphalocele with sterile meals with physiological serum, systemic antibiotic therapy (Shad, J, 2012). EctopiaCordis was first observed 5000 years ago (Taussig HB. ,1982). It has been associated with trisomy 18 and Turner syndrome (King CR ,1980),(Soper SP et al,1986,),(Say B and Wilsey CE. ,1978). Depending on the location, it can be classified into 5 types: cervical (5%), cervical-thoracic and thoracic (65%) and abdominal (10%) (Anderson RH et al ,1987,). Theories about this malformation involve rupture in the early embryogenesis of the corion and the Yolk sac (Kaplan LC, et al ,1985,), (Bieber FR ,1984,) Also, ectopiacordis has also been associated with the administration of intrauterine drugs, results obtained on animal experiments (Jaffee OC, and Jaffee AL. ,1990), (Russo R, et al. ,1993,). The first attempt to treat ectopiacordis was made in 1925 by Cutler and Wilens (Shad, J., et al ,2012,). The first successful repair was made by Koop in 1975 (Saxena AK ,2005).

MATERIAL AND METHOD

We have analyzed the possible causes of a rare malformation called ectopiacordis. Ectopiacordis is a very rare congenital anomaly with a prevalence of 5-8 cases per 1 million live births and is manifested by the absence of anterior chest wall closure, the cord being located outside the chest. Sometimes the ectopic heart can be completely covered by a serous membrane or skin, sometimes it can only be partially covered. This rare malformation occurs between day 14-18 of intrauterine life, localized to the mesoderm. The main cause is the damage of the mesoderm with the lack of sternum closure and implicitly ectopic position of the heart. EctopiaCordis is the result of the lack of maturation of the midline of the mesoderm and the ventral part of the body wall during embryogenesis.The lesions of the embryonic layers are given by a number of factors called theratogenic factors. These include infectious agents, physical agents, chemicals, hormones, mechanical factors, defective factors. Depending on the affected embryonic layers, different fetal malformations may result. Affecting the ectoderm leads to neural tube defects, spine bifida, microcephaly, and endodermic damage results in abnormalities of the digestive tract, such as the Mekel diverticulum. In addition to the disease, this causes malformations such as ectopiacordis,omphalocele.

RESULTS AND DISCUSSIONS

Considering that there is no theory to explain this malformation, we propose the possibility of exposing mechanical damage to embryonic foliage in early embryogenesis, either by manipulating embryos in the fertilization process or by punching, the biopsy for the diagnosis of genetic diseases. Thus, damage to the mesoderm leads to the prevention of embryogenesis in the normal conditions of the patient, including ectoderm and mesoderm.This malformation has been known since ancient times, and no increase or decrease has been observed throughout history, resulting in a sporadic malformation, probably due to accidental point mutations. In terms of treatment, medical technology progress allows the medical and surgical treatment of this anomaly.Also, prenatal diagnosis due to imaging methods and ultrasound can be done early, so mother and doctor know the risks of pregnancy. This malformation is treated in specialized centers for pediatric surgery, but their rarity does not allow very rapid development of surgical techniques, experimental methods are still widely applied and depend on the surgeon's inventiveness and training.

CONCLUSIONS

1. EctopiaCordis could be caused by mechanical damage to the embryo early in the embryogenesis phase.
2. We propose the hypothesis that this malformation is due to the damage of two embryonic leaves and not just the mesoderm
3. It is a known malformation from the earliest times with sporadic occurrence throughout history without changes in the frequency of occurrence.
4. Some studies associate it with genetic diseases such as trisomy and translocation.
5. Pathogenesis theory involves a closure defect in the anterior wall of the trunk.
6. Regarding the treatment of this anomaly, at present, it is possible to intervene effectively because of medical progress, but experimental techniques are still a widely used option.

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