Picture Stories

Ectopia cordis with probable pentalogy of Cantrell

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Introduction

Ectopia cordis is a very uncommon condition, where the heart is located partially or completely outside the thorax[1]. It has an incidence of 8 per million births[2]. In the majority of cases, the heart protrudes outside the chest through a split sternum[1]. However, the ectopic heart can also be located in the neck or abdomen[1].

Case report

A term baby with a birth weight of 2.25kg was admitted to the Sick Newborn Care Unit at the age of 1 hour with the heart completely outside the thoracic cavity (Figure 1).

Mother was not a booked case and no prenatal screening ultrasonography was available. On admission, respiratory rate was 60/min, heart rate was 160/min and oxygen saturation was 85%. Baby also had an omphalocoele, sternal cleft, diaphragmatic hernia and absent diaphragmatic pericardium. However, echocardiography could not be done to detect any intra-cardiac defect.

Discussion

Ectopia cordis is often associated with cardiac anomalies such as atrial septal defect, ventricular septal defect, Fallot's tetralogy and tricuspid atresia[1]. Ectopic heart is due to a failure of proper formation of midline mesoderm during embryonic life. Thus the heart is not protected by pericardium[2]. It may be associated with non-cardiac anomalies such as pentalogy of Cantrell or omphalocele[2].

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Figure 1: ***Ectopia cordis***

Pentalogy of Cantrell comprises omphalocele, anterior diaphragmatic hernia, sternal cleft with or without ectopia cordis, absence or defect of the diaphragmatic pericardium and one or more intra-cardiac defects[3]. It has an incidence of less than 1 in 1,000,000. A locus at Xq 25-26 may be associated[3]. Our patient with an ectopic heart had omphalocele, sternal cleft, diaphragmatic hernia and absent diaphragmatic pericardium. However, as prenatal sonography and echocardiography were not available we could not confirm any intra-cardiac defect.

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