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Incomplete pentalogy of cantrell associated with hydrocephalus

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ABSTRACT

An unbooked primigravida delivered vaginally a female neonate with ectopia cordis, supraumbilical abdominal defect, and sternum defect that is consistent with incomplete pentalogy of cantrell. The neonate was also having central nervous system malformation in the form of hydrocephalus. Neonate expired within 30 min of birth.

Keywords: Ectopia cordis, hydrocephalus, pentalogy of cantrell

IMAGE

The pentalogy of cantrell was first described in 1958.[1] The pentalogy of cantrell is a rare syndrome with an estimated incidence of 5.5/1 million live births.[2] The hallmark of this syndrome is an omphalocele associated with ectopia cordis.[3] The full spectrum consists of five anomalies: A deficiency of anterior diaphragm, a midline supraumbilical abdominal wall defect, a defect in the diaphragmatic pericardium, various congenital intracardiac abnormalities, and a defect of the lower sternum.[1] Based on review of 61 cases of pentalogy of cantrell, toyama suggested the following classification for the syndrome: Class 1, certain diagnosis with all five defects present; Class 2, probable diagnosis, with four defects (including intracardiac and ventral abdominal wall abnormalities) present; and Class 3, incomplete, with various combinations of defects presents (but always including a sternal abnormality).[4] Very few cases of reported pentalogy of cantrell are associated with craniofacial malformation, which include anencephaly, meningocele, cephalocele, hydrocephaly, and exencephaly.

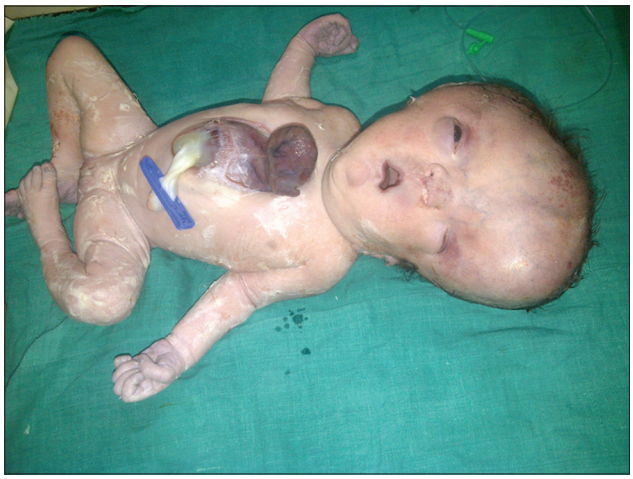

Figure 1: Showing pentalogy of cantrell

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