Clinical Image

Pentalogy of Cantrell or Cantrell Syndrome

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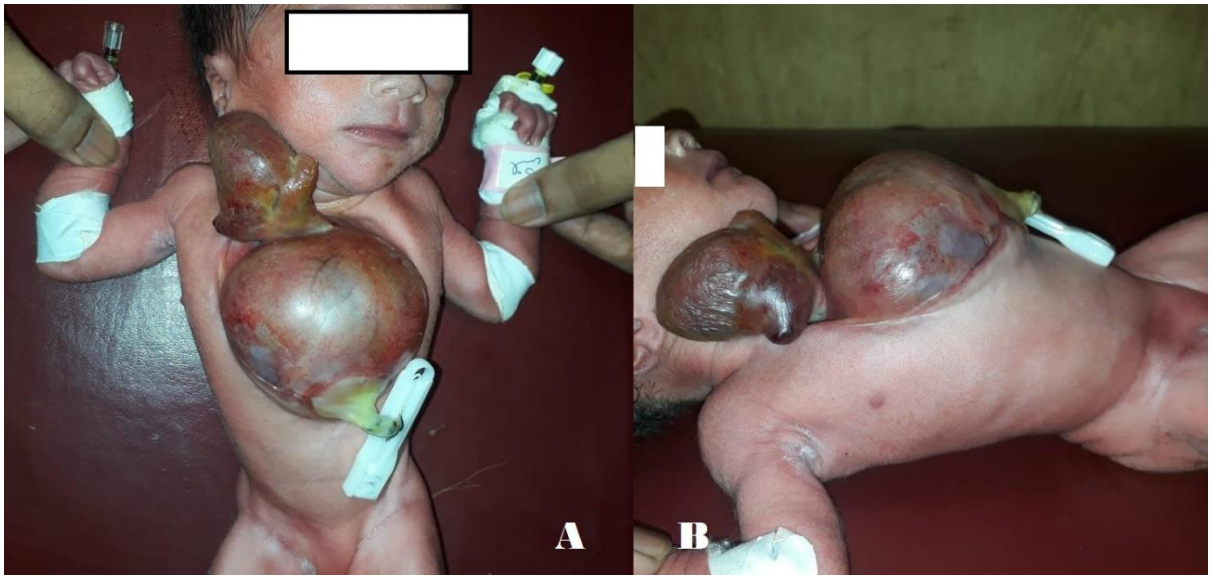

Figure 1: A) Ectopia Cordis and giant omphalocele. B) Lateral view

A 12 hours female neonate, referred to us with heart lying outside the chest and an omphalocele as well. Baby had complete thoracic ectopia cordis, midline anterior thoraco-abdominal wall defect through which liver was protruding, covered by a membrane. Lower sternum was not palpable. Echocardiography showed ASD, VSD, pulmonary and infundibular stenosis.

Pentalogy of Cantrell (PC) is a rare congenital malformation with great variation. Etiology is unknown but defective development of lateral mesoderm with poor differentiation, proliferation and migration during 14-18 days of embryonic life is suggested to be responsible. PC has 3 types, Class I has all the five defects (Omphalocele, Sternal defect, Diaphragmatic defect, Pericardial defect, Cardiac defect). Class II has four defects with intracardiac and abdominal defects as essential components. Class III is incomplete disease expression having combination of defects with sternal defect as an essential component. Early diagnosis with antenatal ultrasound is possible.[1-4] Ectopia cordis can be partial or complete, its location may be cervical, thoracic, thoraco-abdominal and abdominal. Management of PC is challenging and multidisciplinary. Aim is to have a complete survey of other congenital anomalies and closure of defects as early as possible. Overall prognosis is poor and depends on type of PC, position of ectopia cordis and intracardiac defects.[1-4]

Consent: Authors declared that they have taken informed written consent, for publication of this report along with clinical photographs/material, from the legal guardian of the patient with an under-standing

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that every effort will be made to conceal the identity of the patient however it cannot be guaranteed.

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