

SPOT DIAGNOSIS (IMAGE GALLERY)

**BEATING HEART OUTSIDE THE CHEST****Kalyanshettar SS, Navadgi AS, Naganoor RG, Talikoti AB***Navajeevan Children's Hospital, Bijapur, India.*

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A newborn baby presenting with pulsatile mass in the upper abdomen. Examination revealed exomphalos major with part of cardia in it. X ray showed absence of lower sternum with herniation of heart in to the sac of exomphalos. Echocardiography revealed Tetralogy of Fallot. Antenatal sonogram (level 1) done at a peripheral centre in the second trimester was reported as normal.

What is the diagnosis?

Pentology of Cantrell. It is a rare syndrome of congenital malformation of the left pericardium, the heart, the sternum, the diaphragm and the abdominal wall. (1) Usually it presents with exomphalos major with heart and abdominal organs within it. The condition is potentially lethal and should be managed by surgical correction of the defect. The other three defects of the pentology are disruption of all the interposing structures viz. the distal sternum, anterior diaphragm and diaphragmatic pericardium. (2) Although specific etiology is not known, an event occurring prior to differentiation of mesoderm into sternum and layers of pericardium could produce the defect. The timing of the event or insult would be between 14 to 18 days after conception. (2) The complete form may not be expressed always. Incomplete variant forms having three to four of the features have been described. The most common intracardiac defects are atrial septal defect, ventricular septal defect and tetralogy of Fallot. (3) The syndrome can be diagnosed prenatally by careful sonographic examination. As described in the present case, the prenatal diagnosis was missed as it was a level 1 scan. The condition has to be differentiated from isolated ectopia cordis, isolated abdominal wall defect and amniotic band syndrome. The condition carries poor prognosis for survival. Survival of 20 percent was reported by Toyoma et al (4) which included incomplete expressions and mild defects. Others have reported a survival report of 0 percent. (5) Few authors have reported successful repair of the condition. (6).

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