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Radiology

PENTALOGY OF CANTRELL: AN ANTENATAL DIAGNOSIS ON ULTRASONOGRAPHY

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ABSTRACT Pentalogy of Cantrell is a rare disorder characterized by anterior diaphragmatic defect, ventral abdominal wall defect, pericardial defect, intracardiac anomalies and lower sternal defect.[1] Diagnosis of the complete syndrome is made when above five criteria given by Cantrell is fulfilled but incomplete variant forms also exists, here we are presenting a case of pentalogy of Cantrell with incomplete expression of syndrome i.e. Toyoma class III. Our case also showed Exencephaly which is acrania with a disorganized mass of brain tissue.

KEYWORDS : Pentology of Cantrell,Ectopia cordis,Ompalocele, Sternal cleft

I. Introduction: Abnormal formation and migration of the ventral mesoderm and failure of fusion of the transverse septum of the diaphragm and lateral folds of the thorax is postulated to cause Pentalogy of Cantrell[2]. Different types of Cardiac defects are associated which includes septal defects, tetralogy of Fallot, left ventricular diverticulum, and Ebstein malformation [3]. Other associations are cleft lip or palate and exencephaly, encephalocele, spina bifida, hydrocephalus and clubfoot, absent radius anomalies [4–7], chromosomal abnormalities e.g. trisomy 18, cystic hygroma ,malrotation of the colon, single umbilical artery, dysplastic kidney, gastroschisis, diastasis recti, absent gall bladder [8,7].

II. **Case report**: A 22 year old Gravida 3 Para 2 woman came for routine antenatal ultrasound during second trimester with history of amenorrhea since 6 months. Her 1st baby is healthy male of 3.5year born by normal FullTerm Vaginal delivery,her 2nd baby is female born by normal FullTerm Vaginal delivery,baby died at age of 15 monts cause was not known to mother,no history of consanguineous marriage, no family history of congenital anomalies, no history of intake of teratogens, Nohistory of Rh incompatibility was present. Blood pressure was normal with normal blood glucose. Her routine hematological and biochemical profile was normal. On usg single live unstable fetus, of age 23weeks of gestation was present. Amniotic fluid index was 27 suggestive of Polyhydramnios. Exencephaly-which is acrania with disorgainised brain tissue mass was found as cause of polyhydromnios as evident by frog eye sign.Large omphalocele noted containing liver,gallbladder,stomach,spleen,stomach,intestines, ,fetal heart was present outside thorax protruding in to sac though sternal defect suggestive of ectopia cordis(thoraco-abdominal type) ,however no evidence of intracardiac abnormalities was evident that's why we kept this diagnosis as Toyoma class III that is incomplete expression of syndrome. All the limb bones was normally formed with nomal usg apperence of bony spine was also noted.

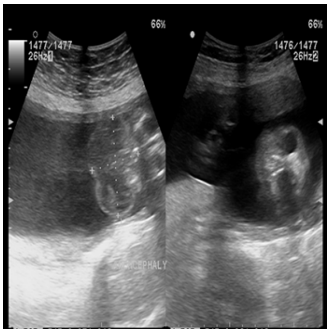

Fig 1 shows absence of normally formed skin, cranial bones, and brain superior to the orbits – frog eye sign. calipers shows disorgainised mass of brain tissue

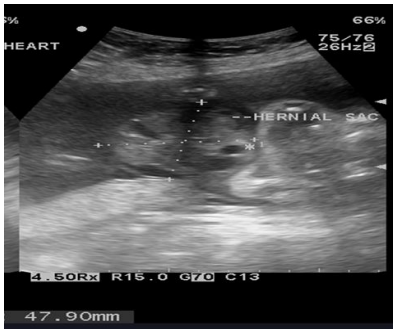

Figure 2 shows. Herniated sac covered by membrane,containing gallbladder & stomach

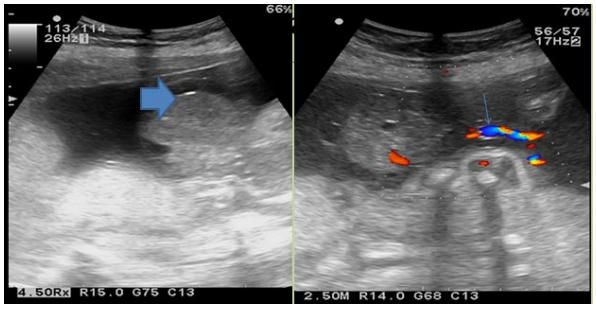

Fig 3- Herniation of liver and intestines thick arrow and thin arrow shows Cord seen attached to apex of sac

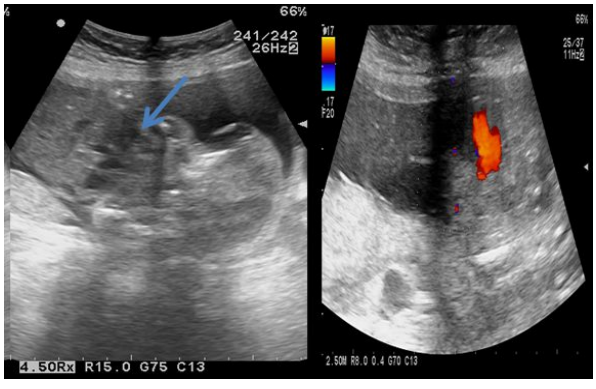

Fig 4-shows Heart protruding out of thorax into the sac

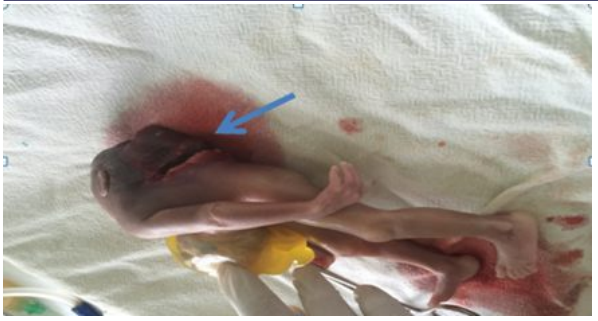

Fig 5- Baby delivered by elective caesarian section,Exencephaly noted. All extremities of abortus was normal no defect noted.

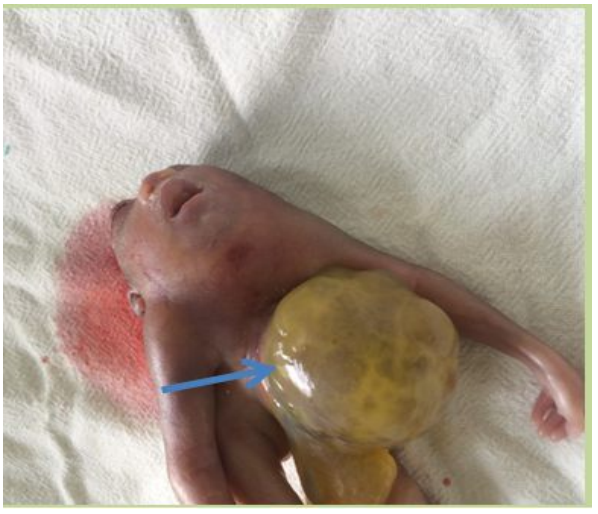

Fig.6 large omphalocele with defect in sternum

III. Discussion& Conclusion

Reported prevalence of Pentalogy of Cantrell is 5.5 cases per 1 million live births [4]. In Most cases risk for recurrence is negligible . Omphalocele accounts for 63% of reported cases [3]. Presence of omphalocele of the upper abdominal wall with ectopia cordis is sufficient to suggest the diagnosis of Pentalogy of Cantrell .Toyama [10] classified the pentalogy of Cantrell as class I, definite diagnosis i.e. all five defects are present ,class II, probable diagnosis, in this class only four defects are present, including intracardiac and ventral wall abnormalities; and class III i.e. incomplete expression, in which various combinations of defects present, including a sternal defect. Differential diagnosis includes OEIS complex i.e. Omphalocele, exstrophy of the bladder ,imperforate anus & spinal defects however presence of normal fetal heart helps to differentiate it from pentalogy of cantrell.Body stalk anomaly may also pose diagnostic challenge because it shares common feature of omphalocele and variable ectopia cordis but presence of amniotic band, fixed position of fetus,limb defects helps to differentiate it from pentalogy of Cantrell (see table 1 for further differentiating feature),cases with omphalocele together with sternal malformations must be evaluated for other components of the Cantrell anamoly.

Table 1

	Pentalogy of cantrell	**OEIS complex**	**Body stalk anomaly**
Abdominal wall defect	Midline supraumbilical	Midline infraumbilical	Large,eccentric, left lateral more common
Fetal position	Not fixed	Not fixed	Fixed& fetal part may adhere partly with placenta
Umbilical cord	Normal	normal	Short / absent[11]
Limb defect	Less common	Present occasionally	common
Fluid filled urinary bladder	Present	Persistently absent	present

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