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Is Mermaid a Curse….A Rare Entity?

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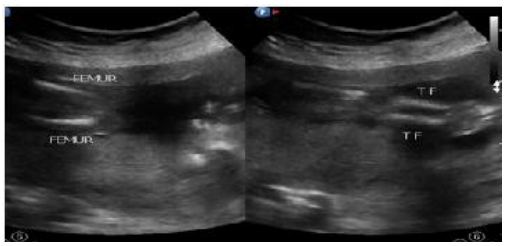
Introduction: *The mermaid of Greek and Roman mythology, was depicted as having the head and upper body of a human and the tail of a fish. It was considered to be one of the most beautiful creatures that ever lived but in truth, the mermaid baby also known as syrinomelia is almost always lethal. It is a severe form of caudal regression syndrome that results in a fusion of the lower extremities, which is not compatible with life. A spectrum of anomalies affects primarily the musculoskeletal, genitourinary, cardiovascular and gastrointestinal systems. Sirenomelia is a rare congenital malformation with an incidence of 1.5 - 4.2 per 100,000 births. It has a relative risk of 200-250 in diabetic pregnancies*
Keywords: *Sirenomelia , caudal regression syndrome, congenital anamoly, diabetes mellitus*

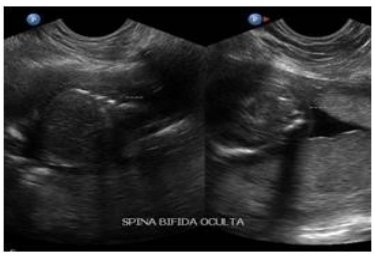
I. Case Report

A 30 year old Gravida-5, para-2, living-1, abortion-2, with 23 weeks of gestation came to our hospital for regular antenatal check up when she was diagnosed to have multiple congenital anamolies on scan. Her obstetric history indicated that the first pregnancy ended up as IUD, the baby was found to have limb anamoly.2nd and 3rd pregnancies were terminated as the fetuses were found to have growth retardation. She has one female healthy baby.

The present pregnancy is the fifth pregnancy. The first trimester was uneventful . She has taken regular folic acid supplementation. She is not a known case of diabetes mellitus. There was no family history of diabetes mellitus. On investigation, her blood glucose levels were found to be within normal limits

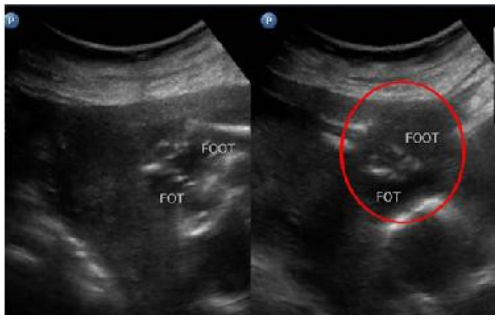


Both fused femur ,tibia and fibula on ides.

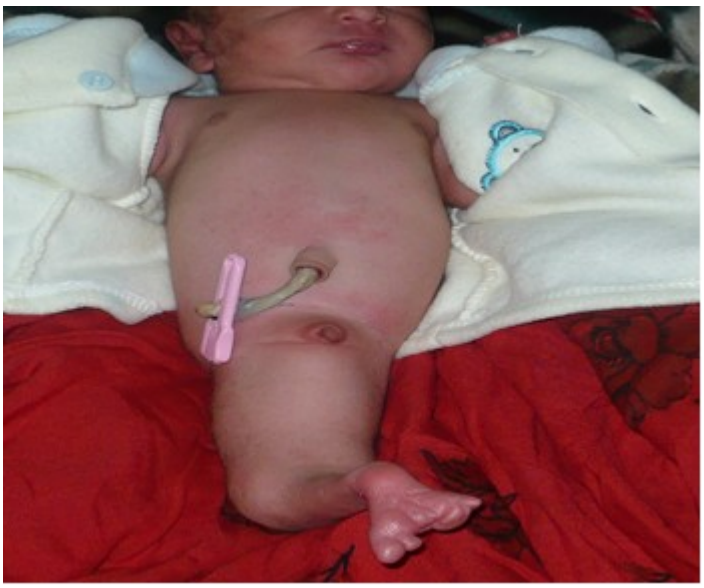


Renal agenesis present on both sides both

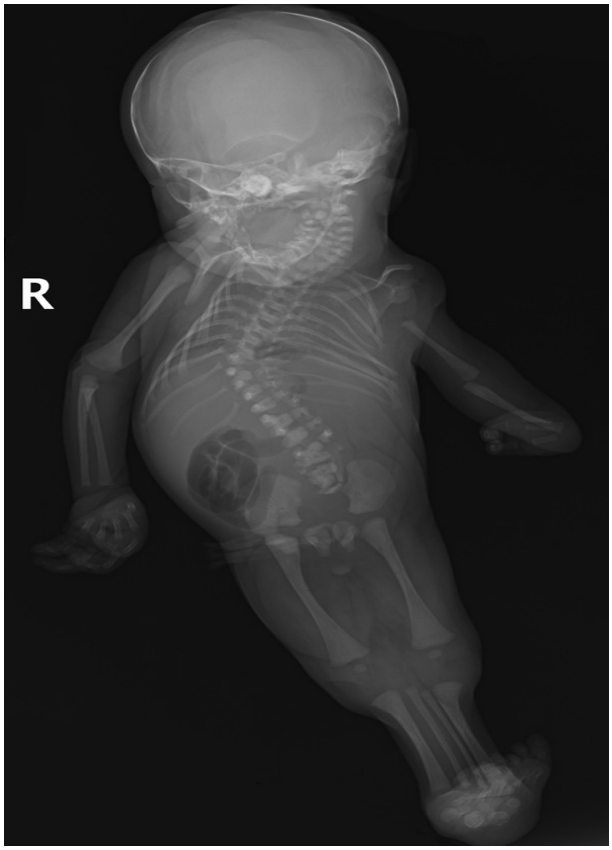
Both feet are visible.



The couple was counseled regarding the prognosis and that the anomalies were incompatible with life. The couple opted for termination of the baby and it was done by prostaglandin E1(misoprostol) induction .



A 450 gm live baby was delivered. No active resuscitation was done. Placenta weighed 200gm. Umbilical cord had 1 artery and 1 vein. Liquor is scanty in amount. The baby had low set ears, absence of external genitalia, atresia of anal canal, lower limbs not well formed appear to be fused with both feet separate On autopsy an anterior abdominal wall defect, lung hypoplasia were present



II. Discussion:

Etiology of sirenomelia is uncertain.

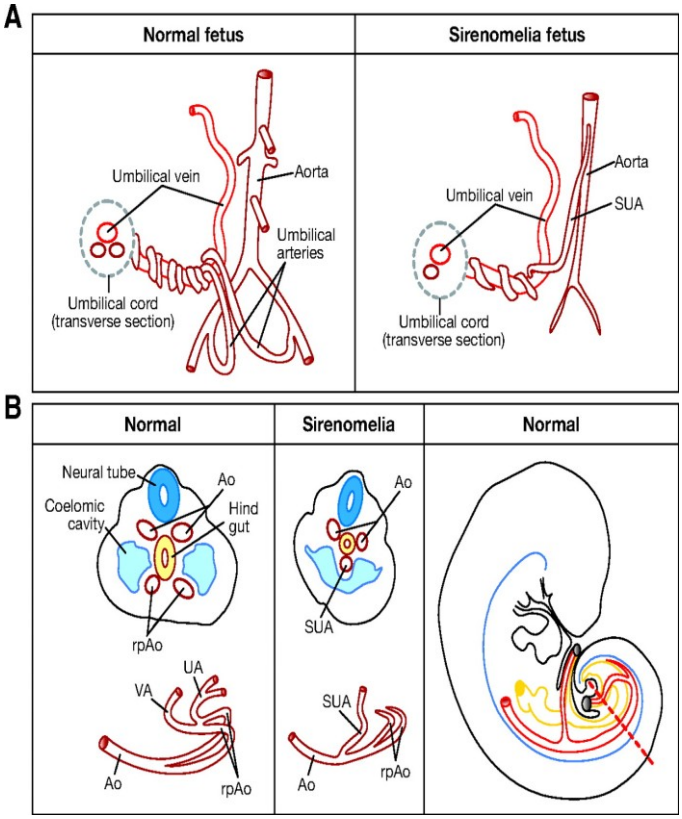
1. Abnormality in blastogenesis, and it is due to disturbances during blastogenesis (days 1 to 28) which represents the primary field. The pattern of single or multiple malformations occurring in association with lower vertebral column maldevelopment is governed by perturbations in the morphogenic pathways

2. VASCULAR STEAL PHENOMENON: This causes severe ischaemia of caudal portion of fetus. Here an aberrant vessel derived from the vitelline artery shunts blood from high abdominal aorta directly the umbilical cord to placenta . This vessel in effect, acts as an umbilical artery. The result is severe hypoperfusion of structure distal to the origin of abnormal vessel, as this vessel ―steals blood from caudal portion of fetus.

3. Altered oxidative metabolism from maternal diabetes may cause production of free oxygen radicals in the developing embryo, which may be teratogenic.

4. Genetic aspects of sirenomelia : Gain of function of retinoic acid(RA) signaling or loss of function of bone morphogenetic protein (Bmp) signaling is seen in CRS.

5. Animal experiments have shown that CRS-like syndrome could be induced by agents including retinoic acid, diethylpropion, lithium, sulfamide, cadmium, lead, ochratoxin A, vitamin A deficiency, radiation, hyperthermia, organic fat solvents, and 6-aminonicotinamide.



III. Conclusion :

-It can be diagnosed as early as 13 weeks of pregnancy. Third trimester ultrasound is impaired by severe oligohydramnios where as it is easier to diagnose it in 2nd trimester.

- Patients who present with oligoydramnios along with growth retardation either with or without history of leaking, we should look for fetal kidneys and fetal bladder along with fetal vasculature.

- Therapeutic abortion can be carried out in earlier gestation due to invariably lethal condition, because of bilateral renal agenesis which is associated with severe oligohydramnios , lung hypoplasia and growth retardation.

-If diagnosed early, termination can be offered, which is less traumatic both physically and mentally to mother.

-The maternal glucose levels should be optimal in preconceptional period and 1st trimester to prevent this condition.

- Few surviving patients need a multidisciplinary approach of treatment

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