MERMAID BABY WITH MULTIPLE CONGENITAL ANOMALIES IN MONOCHORIONIC TWIN GESTATION IN DIABETIC MOTHER

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ABSTRACT: Sirenomelia, also known as sirenomelia sequence, is a severe malformation of the lower body characterized by fusion of the legs and a variable combination of visceral abnormalities. Infants with cyclopia or sirenomelia are born at an approximate rate of 1 in 1,100,000 births. We are presenting a report of a rare case of Mermaid baby with multiple congenital anomalies in monochorionic twin gestation in a diabetic mother.

KEY WORDS: Mermaid, Diabetes, Monochorionic twins, congenital anomalies.

CASE REPORT: A 22 year old Gravida 2 Para 1 Living 1, with previous LSCS, unbooked, got admitted to our obstetric ward at 33weeks+3days of gestational age. She was a known case of overt Diabetes mellitus on irregular treatment. On admission her blood sugar levels were high(FBS-248mg%, PPBS-480mg%) .She was started on Insulin. Ultrasonography revealed monochorionic diamniotic twin gestation corresponding to 33weeks+3days with gross polyhydramnios in one sac and no evidence of congenital anomalies were noted. A course of corticosteroids was administered and elective caesarian was performed at 34 weeks of gestation.

Twin 1 –1.5kg,preterm,male baby, delivered as breech showed evidence of Mermaid anomaly, neural tube defect(spina bifida occulta -lumbar region), single kidney, single ventricle, imperforate anus, single umbilical artery, sacral agenesis. Baby died after 2 hours.

Twin 2 –1.7kg, preterm, male baby, delivered as cephalic, no obvious congenital anomalies were detected.

Both mother and twin 2 were healthy and discharged on 16th day.

DISCUSSION: Sirenomelia, also known as sirenomelia sequence, is a severe malformation of the lower body characterized by fusion of the legs and a variable combination of visceral abnormalities[1]. Clinical studies of sirenomelia have given rise to two main pathogenic hypotheses. The first hypothesis, based on the aberrant abdominal and umbilical vascular pattern of affected individuals, postulates a primary vascular defect that leaves the caudal part of the embryo hypoperfused. The second hypothesis, based on the overall malformation of the

caudal body, postulates a primary defect in the generation of the mesoderm. Infants with cyclopia or sirenomelia are born at an approximate rate of 1 in 100 000 births[2,3].

Eight malformation monitoring systems around the world jointly studied the epidemiology of these rare malformations: 102 infants with cyclopia, 96 with sirenomelia, and one with both conditions were identified among nearly 10.1 million births. Maternal age is somewhat increased for cyclopia, indicating the likely inclusion of some chromosomally abnormal infants which were not identified. About half of the infants are stillborn. There is a female excess among infants with cyclopia. Excess twinning occurred for cyclopia and possibly also for sirenomelia[4]

In Greek mythology, the Sirens were three creatures with the head of a woman and the body of a bird from the wings down. They were dangerous to sailors, whom they narcotized with their enchanting music and voices to later kill them, despite the present perception of Sirens as romantic and cute creatures, the sirenomelia human malformation is a severe condition[1].

The diagnosis, which is obvious at birth, is currently performed by prenatal ultrasonography. Antenatal ultrasonography clues include oligohydramnios, renal agenesis and a fibula positioned between the tibiae[5]

Although the primary molecular defect underlying sirenomelia remains unknown,the two main pathogenic hypotheses are the vascular steal hypothesis and the defective blastogenesis hypothesis[6].In humans, a genetic etiology for congenital caudal anomalies has only been confirmed for the Currarino syndrome, a sacral agenesis caused by mutations in the homeobox-containing gene *HLXB9*[1] . By contrast, to date, all reported incidences of sirenomelia in humans have been sporadic cases. Maternal diabetes has been associated with both caudal regression syndrome and sirenomelia[7]

Although it is possible that, in humans, sirenomelia is an autosomal-dominant genetic condition, it has a combined genetic and environmental component maternal diabetes and heavy metals have been described as important environmental risk factors for caudal malformations[1] .

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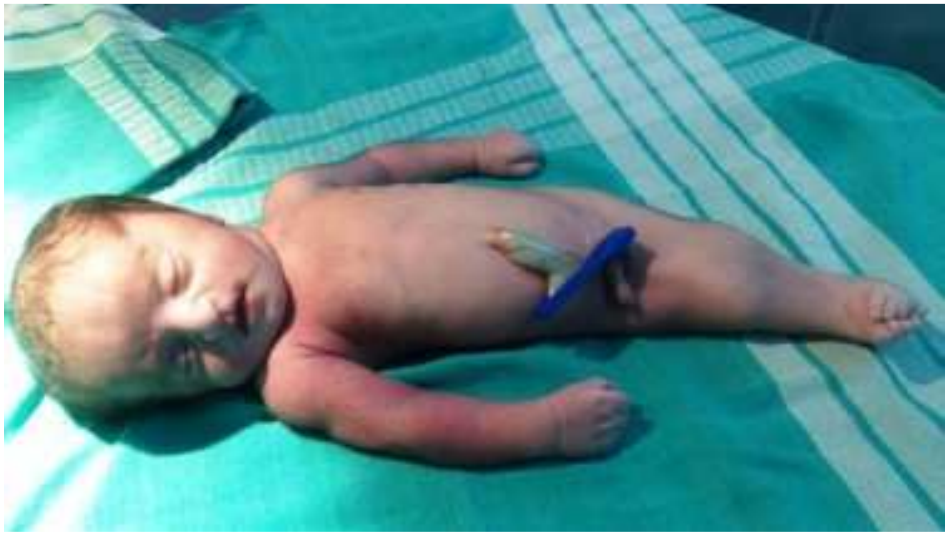

Figure 1 : Mermaid Anomaly

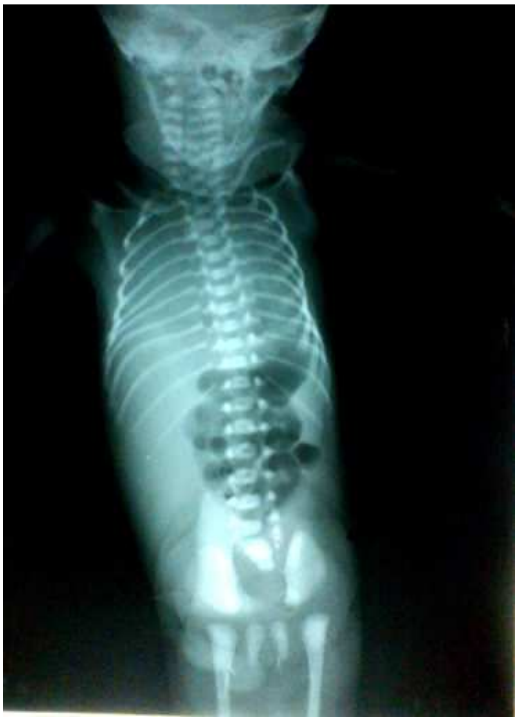

Figure 2 : Sacral Agenesis

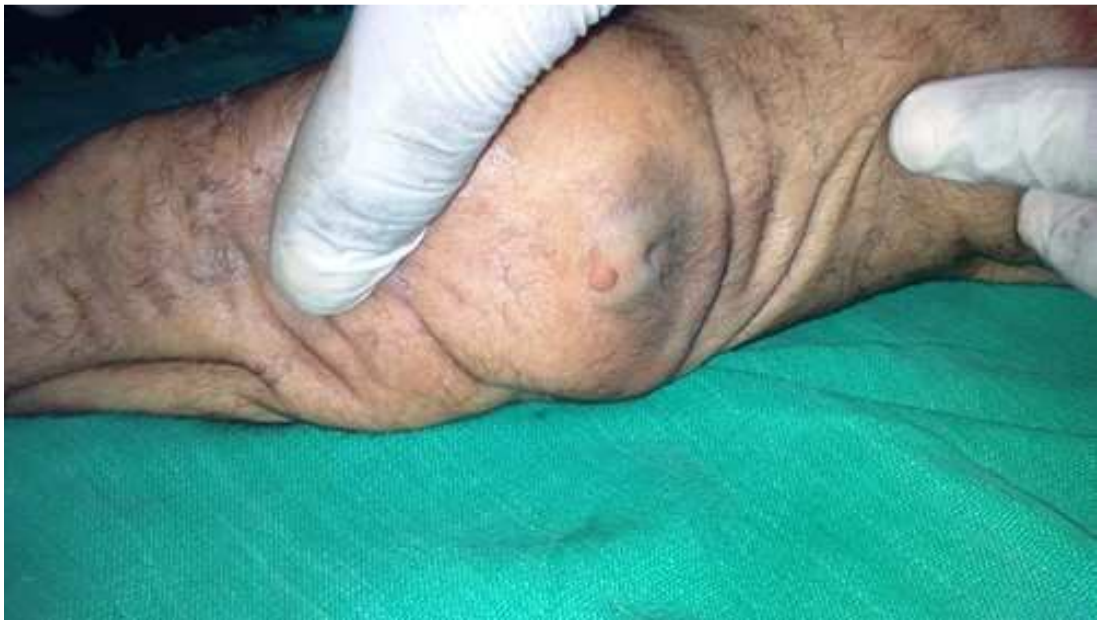

Figure 3 : Spina Bifida Occulta