

CASE REPORT

SIRENOMELIA: THE MERMAID BABY - RARE CASE REPORT

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ABSTRACT: Sirenomelia "THE MERMAID SYNDROME" condition is found approximately. 1:1,00,000 live births¹ and is fatal within the first day because of complication of abnormal kidney and urinary system development. Sirenomelia is characterised by fusion of the lower limbs with absent kidneys, absent external genitalia and other gastrointestinal defects. It results from the failure of normal vascular supply from the lower aorta in vitro, maternal Diabetes mellitus has been associated with caudal regression syndrome and sirenomelia. We are reporting rare case of mermaid baby which was delivered in our hospital. Early diagnosis by ultrasound scan and termination should be our goal.

KEY WORDS: Mermaid syndrome, Sirenomelia.

INTRODUCTION: MERMAID SYNDROME was first described by Rocheus in 1542 and Palfyn in 1553 and called after the mythical Greek sirens.^{2,3} This lethal anomaly is associated with a number of genitourinary and anorectal defects³⁻⁴. This syndrome has strong association with maternal Diabetes mellitus with relative risk 1;200-250 and 22% of foetuses will have diabetic mother^{5,6}. Intake of haloperidol antenatally is considered a cause for Sirenomelia. Other proposed teratogens are cadmium, lead, vitamin A. Nutritional deficit, vascular hypoperfusion has also been proposed as a possible etiological factors.

CASE REPORT: A 30 year old woman delivered a live baby at 34 wks of gestation with fused lower limbs. The baby didn't cry immediately after birth and required resuscitation in the form of bag and tube ventilation. After discussing with parents and as per their request not to resuscitate baby was kept under hood oxygen. The neonate did not survive this lethal abnormality and died after 4 and half hours of life. There was history of ingestion of tobacco leaves by the mother during antenatal period. There was no history of ingestion of other teratogenic agents or diabetes mellitus.

X-rays revealed lower limbs were fused completely into single limb and one femur and one tibia were seen.

Immediately after birth, Ultrasound abdomen showed bilateral agenesis of kidney and urinary bladder. Echocardiography showed large Atrial Septal Defect.

DISCUSSION: The diagnosis of sirenomelia by ultrasound is done by the presence of fused femur and decreased distance between two femurs and decreased or absent mobility of two lower limbs, and associated with skeletal and lumbar spine deformities^{2,5}. In addition bilateral renal agenesis, heart and abdominal wall defects are seen on scan⁵. Diagnosis is done in second trimester as oligohydramnios as the alerting sign⁷. Sirenomelia can be differentiated from

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caudal regression syndrome with presence of “persistent vitelline artery”^{1,8} and the syndrome is not associated with any chromosomal abnormality⁶.

The condition is seen in in 100-150 times more frequently in monozygotic than dizygotic twin or singeltons and males are three times more affected than females ².

Although sirenomelia has been described as a rare lethal anomaly, 9 mermaid syndrome cases have been reported surviving after reconstructive surgery ⁹. The most important characteristics of survival of the affected newborn is the presence of functional kidney.

There are three different variant depending on the degree of fusion of the lower limbs.

- 1) Symeliaapus: no feet are present and the limbs are totally fused into a single limb; 1 femur and 1 tibia is present.
- 2) Symeliaunipus: one foot is present (a partial fusion of both feet), 2 femur, 2 tibia, 2 fibula⁹are present.
- 3) Symeliadipus: feets are present giving the appearance of fins, hence the term ‘mermaid fetus’ for this condition. The fusion of the limbs extends only till the ankle.

CONCLUSION: SIRENOMELIA is fatal congenital anomaly and has a strong association with maternal diabetes, early diagnosis and termination should be our goal. In addition a second ultrasound should be performed after 4-6 weeks after the initial first trimester (8-9 weeks) scan. So that gross structural anomaly can be picked up and termination of pregnancy can be planned.

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Mermaid Baby



X-ray of Mermaid Baby

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