

Case Report

Mermaid and its Association with Single Umbilical Artery: Review of Literature

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ABSTRACT

Sirenomelia or Mermaids syndrome is a rare anomaly.

The exact cause for sirenomelia is yet to be known. Multiple risk factors and theories for its development have been proposed. In this article we have reviewed the association between single umbilical artery and sirenomelia.

Key Words: Sirenomelia, caudal regression syndrome, mermaid.

INTRODUCTION

Mermaid or Sirenomelia is a congenital structural anomaly characterized by an abnormal development of the caudal region of the body with different degrees of fusion of the lower extremities. It bears resemblance to the mermaid of Greek mythology. The fetal legs are fused together giving the appearance of a mermaid hence also called as Mermaid deformity. This deformity is also known as Symmelia, Symposia, Sympus, Uromelia and Monopodia Sirenomelia is a very rare congenital anomaly seen in approximately 1 in 1,00,00 live births. ^[1]

Ano-rectal, genito-urinary, cardiac anomalies are also seen to be associated with sirenomelia. We report a rare case of sirenomelia of 22 weeks of gestation with a single umbilical artery suggesting its association.

CASE REPORT

A 24year old G3P1+1L1 with previous caesarean delivery 2 years back

wanted termination of pregnancy due to ultrasound findings suggestive of 22 weeks pregnancy with fetal polycystic kidneys, congenital heart disease and with absent liquor .There were no medical or obstetric risk factors. There was no personal or family history for Diabetes Mellitus or congenital abnormality. Her previous pregnancy was also uneventful. The couple was counseled and they opted for termination of pregnancy and were admitted. Medical termination tried but failed. Patient was taken up for hysterotomy in view of failed induction. A 400 gm dead fetus extracted with multiple anomalies (**Fig 1**): Both lower limbs fused (**Fig 2**), polydactyl, not well formed ears, anal atresia, sex not well made out. Placenta and cord sent for histopathology - single umbilical artery. Autopsy of the fetus could not be done as the patient and her relatives denied. Patient postoperative period was uneventful and she was discharged on sixth post operative day in stable condition.



Figure 1: Sirenomelia / Mermaid fetus



Figure 2: Mermaid Fetus with limb deformity

DISCUSSION

In the world literature around 300 cases of sirenomelia have been reported. [2] (Table 2). The diagnostic triad suggested by Raabe et al for Sirenomelia / mermaids syndrome consists of (i) fused lower extremities (ii) bilateral renal agenesis (iii) oligohydramnios. [3] Caudal regression syndrome, sirenomelia sequence and VACTERAL anomalies have similar presentations. Initially sirenomelia was considered as severe form of caudal regression syndrome but now it is considered as a separate entity. One of the risk factor associated with it is maternal diabetes. It is seen in 2 % of sirenomelia cases and seen more frequently- 22% in those fetuses born with caudal regression syndrome. Teratogens like cadmium, retinoic acid and cyclophosphamide and genetic factors also seem to play a role in its occurrence. [4]

Various theories for its pathogenesis have been known: [4]

1. Pressure theory by Ballantyne: Intrauterine pressure? Amniotic fluid pressure may act on the caudal end of embryo causing the deformity
2. Primary Failure by Bolk: Failure of development of somites
3. Nutritional theory by Weigert: Hypoplasia of vasculature supplying the caudal end

4. Caudal mesoderm injury by Davis et al: Injury between 28-32 days causes developmental arrest of primitive streak and thus the midline cloacal and urogenital structure are destroyed
5. Neural tube overdistension by Gardner and Brenner: it leads to tubal roof plate expansion and lateral rotation of mesoderm by 180 degree causing fusion of lower limb buds
6. Vascular steal theory by Stevenson et al: Single umbilical artery and malformed vessels arising from aorta below this aberrant vessel leading to decrease in blood supply to the caudal portion.

A direct relationship between single umbilical artery and sirenomelia has been seen. Umbilical cord consists of 2 arteries and one vein. Single umbilical artery is a condition in which the cord has only one artery and one vein. It is seen in 0.5 to 6% of pregnancies. It is seen to be associated with multiple congenital and chromosomal abnormalities. The yolk sac and the body stalk fuse to form umbilical cord between 3 to 5 weeks of gestation. Two umbilical veins fuse to form a single vein which carries oxygenated blood from the placenta to the fetus. The umbilical arteries arise from the allantois, which is a portion of the yolk sac which carries deoxygenated blood from the fetus to the placenta. One of the umbilical arteries may undergo primary agenesis or persistence of

the original single allantoic artery of the body stalk, and secondary atresia of a previously normal umbilical artery could result in single umbilical artery. The latter mechanism is the most common. [5] Types of umbilical artery agenesis and the abnormalities associated with it have been

described in Table 1. Single umbilical artery and sirenomelia are seen to be associated together. A review of literature about their association and other anomalies seen in sirenomelia has been discussed in Table 2.

Table1: Severity of umbilical artery agenesis and abnormalities associated with it : [5]

Type of umbilical artery agenesis	Frequency	Origin of Single umbilical artery	Umbilical Vein	Number of cord Vessels	Associated Abnormalities
I	98%	Allantoic from left or right Iliac artery	Left	2 One artery One vein	CNS, Lower Gut Acardia Short cord Syndrome
II	1.5%	Vitelline from SMA	Left	2 One artery One vein	Severe malformation Sirenomelia Caudal regression Anal agenesis
III	Very rare	Vitelline or allantoic Iliac artery or SMA	Left and persistent right	3 One artery Two veins	TAPVD Renal agenesis Ipsilateral limb reduction Unicornuate uterus Hydranencephaly
IV	Very rare	Vitelline or allantoic Iliac artery or SMA	Right	2 One artery One vein	Embryos lost before prenatal or pathological assessment

Table 2: Review of literature of sirenomelia cases with single umbilical artery: [1,2,6-8]

Authors	Number of cases	Associated anomalies
Monteagudo et al	Two cases: first trimester	Increased nuchal translucency, large abdominal vessels, single umbilical artery
Patel and Suchet	2 cases - 19 weeks 1 case - 18 weeks	Sacral dysplasia, single femur, anorectal atresia, blind-ending colon, renal agenesis, absence of renal arteries, single umbilical artery , no external genitalia, anhydramnios
Browne et al	?	Oligohydramnios, absent lower limb, absent bladder, renal agenesis, a malformed pelvis, scoliosis, a single umbilical artery
Horikoshi et al	Ist case - 24 weeks IInd case - twin pregnancy 24 weeks	Oligohydramnios, renal agenesis, no bladder, a single umbilical artery , only one leg visualized
Sepulveda	1 case - 23 weeks	Severe oligohydramnios, small chest, absent bladder, bilateral renal agenesis, a single umbilical artery , absence of renal arteries, no branching of the main abdominal vessel into the normal left and right common iliac arteries
Akbayir et al	1 case- 11 weeks	A single pelvic bone, fusion of the lower extremities, a single umbilical artery , absence of the iliacal branching, uncertainty about the presence of renal tissue
Taori KB et al	1 case - ? weeks, 1.5 kg	Absence of external genitalia, sacral agenesis, spina bifida, B/L absent radii, single umbilical artery , complete fusion of femur
Morfaw FL	1 case- 38 weeks	Distended abdomen, single umbilical artery , undetermined external genitalia, fused lower extremities
Bibhuti B Das MD et al	1 case - 40 weeks	Anal atresia, B/L renal agenesis, single umbilical artery , fused lower limbs with medial position
Marybeth Browne MD et al	1 case - 40 weeks	Absent lower limb, absent bladder and kidneys, malformed pelvis, scoliosis, large cystic mass in lumbosacral region, single umbilical artery

CONCLUSION

Sirenomelia is a rare and fatal congenital anomaly and has association with single umbilical artery. Early prenatal

diagnosis by first trimester scan should be the aim to minimize the trauma related to the termination of pregnancy at advanced gestation. In addition, where possible, a

second US scan should be performed 4-6 weeks after the initial 8-9 weeks scan so that gross structural anomalies are detected and termination of pregnancy be considered earlier. In patients diagnosed of single umbilical artery other congenital and chromosomal abnormalities are to be ruled out including sirenomelia.

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Conflict of Interests

The authors declare that there is no conflict of interests regarding the publication of this paper.

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