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Mermaids of the Land: A case report from a Secondary care hospital in South India.

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Abstract: *Mermaid syndrome also known as Sirenomelia is a rare congenital malformation. We report a case of Sirenomelia in a secondary care hospital in south India. This congenital anomaly is a rare entity which can be diagnosed during antenatal visit. This case report will enable us to understand the syndrome and the role of nurses in counselling and psychological support for such rare cases.*

Keywords: *Mermaid syndrome, Sirenomelia, Caudal regression syndrome*

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I. Introduction

Sirenomelia, the most severe form of caudal regression syndrome, a complex developmental disorder. The term "Sirenomelia" is used from the Greek and Roman mythological stories. It is often associated with severe life-threatening complications and is fatal in the first year of life. A new-born with legs joined together features a mermaid-like appearance (head and trunk like humans and a tail like fish). In most cases they die shortly after birth. The mermaid syndrome (Sirenomelia) is an extremely rare anomaly, an incidence of 1 in 100,000 births, pregnancies[1] and affects males more than females[2].

The exact cause of Sirenomelia is unknown. It is an autosomal dominant disorder resulting from a new spontaneous mutation and is more likely to be caused by a combination of genetic and environmental factors[3]. Any damage to the mesoderm of the embryonic tail between 13 and 22 days may cause lower limb disorders in new-born[4]. These babies are born dead or they die shortly after birth[5]. Retinoic acid (RA), maternal diabetes, and heavy metals in experimental models were an important environmental danger element involved in the development of embryo tail anomalies. RA signaling in embryonic length has an essential role in fetal development, in particular in the caudal area[6].Interference with the RA signaling pathway in the development of Sirenomelia represents its capacity relationship with environmental elements because retinoic acid levels are related to genetic elements, nutrients, and iatrogenic effects[5].

Maternal diabetes is a major risk factor for multiple caudal anomalies during pregnancy[7] and is the only known disease in the mother that is associated with the mermaid syndrome[8,9,10].

Classification

According to Stocker and Heifetz (1987) Sironomelia is classification based on presence of ossified structures[11]. The classification is presented in Table 1.

Table 1
Classification of Sironomelia

Type	Descriptions
I	Presence of two separated femurs, two tibia and two fibulae
II	Presence of two separated femurs, two tibiae and a medially fused fibula
III	Presence of two separated femurs, two tibiae and no fibula
IV	Presence of a partially fused femur, two tibiae and a medially fused fibula
V	Presence of a partially fused femur, two tibiae and no fibula
VI	Presence of a complete fused femur and a single fused tibia
VII	Presence of a complete fused femur

There are 3 unique evidences of abnormalities of the lower extremities in Sirenomelia:

a) Symelia apus: There are no toes and feet, legs are fully conjoined, only femur and a tibia are visible.
b) Unipus symelia: There is a foot (an incomplete mixture of both legs), however there's two femur, two tibial and fibular bones.
c) Symelia dipus: There are leg-like fins and lower limb connection after knee may be visible.
d) Embryologically it's a caudal blastemal defect, because of the persistence of the vitelline artery

Clinical Manifestation

The signs and symptoms of babies with Sironomelia are referred to as the VACTERL association which is a non-random association of birth defects that affects multiple organ systems[12].

- **V**ertebral abnormalities
- **A**nal atresia
- **C**ardiac defects
- **T**racheal anomalies including tracheoesophageal fistula
- **E**sophageal atresia;
- **R**enal and radial abnormalities
- **L**imb abnormalities

Other defects of Sironomelia may include[13, 14]

- Congenital heart defects
- Respiratory complications: Pulmonary hypoplasia.
- Lumbosacral spine abnormalities: Lordosis, Meningomyelocele
- Defects of the abdominal wall: Omphalocele, Absence of the spleen and/or the gallbladder has also been reported.
- Urogenital abnormalities: Renal agenesis, cystic malformation of the kidneys, absent bladder, urethral atresia, imperforate anus,

Management

Mermaid syndrome is fatal in most cases due to pulmonary hypoplasia and renal failure resulting from renal agenesis. Mostly newborns with mermaid syndrome die within the next few days. Very few cases have been reported where a child with Sirenomelia survived. Sirenomelia fetus usually die because of the hypoplastic lungs or from renal failure. The management of the complications associated with this condition proved to be costly and difficult. Managing Sironomelia requires the coordinated efforts of a group of medical specialists. Balloon-like tissue expanders are inserted beneath the pores and skin, it makes the skin stretch and grows. The skin is used to cover the legs as soon as they're separated [14,15]. An early antenatal analysis with the usage of ultrasonography can be considered in medical termination of pregnancy.

The nursing management of mother with a Sirenomelia is discussed using a case report. Mrs. A, 24-year-old female G3A2 at 38+5 weeks of gestation with pre-gestational diabetes mellitus on insulin therapy was identified to have an early onset IUGR and foetus with multiple anomalies. She delivered a single macerated still birth male baby of weight 1.44 kg by assisted breech delivery. The pregnancy was from a third-degree consanguineous marriage. She was compliant with her antenatal consultations and was identified with early onset of IUGR and foetus with multiple anomalies, anhydramnios on morphological scan. She was not exposed to any teratogenic drugs or illnesses. Previous two abortions were spontaneous at 3 months. Her pregnancy was uneventful.

The baby did not cry at birth and the APGAR scores were zero at one minute and five minutes. On physical examination, the baby had fusion of the lower limbs. The umbilical cord consisted of a single artery and one vein. External genitalia were not well distinguished and an imperforate anus was also seen. The scan revealed the anhydramnios, bowed femur and bilateral kidney were not visualized. Foetogram revealed two tibias, unvisualized foot bones, white out appearance of the lungs, absent sacrum and single femur (see Figure 1 & 2).

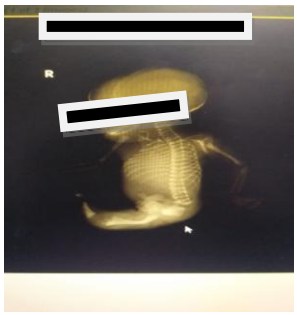

Figure 1. Fetogram

Figure 2. Sirenomelia baby

The mother presented with anxiety and fear. She was unable to accept the death of the fetus and the anomaly. The nursing care of the mother is presented below:

Anxiety related to loss of a new born: Assessed the anxiety stage of the mother, provided spiritual help and diversional therapy. Encouraged her to ventilate her feelings and ensured support from a Counsellor.

Risk for infection related to breast engorgement (no breast feeding) secondary to loss of a new born: Taught her to express the breast milk with the assist of the syringe, applied warm fomentation, advised her to smooth the breast and put on suitable breast assist apparel and to rubdown the breast during bath to prevent breast engorgement

Risk for ineffective child bearing process related to loss of a new born secondary to congenital malformations: Assessed for the support system (family, pals and spouse and children), her physical and emotional desires. Taught her postnatal care (nutrients, infection prevention and prevention of breast engorgement). Showed empathy whilst communicating with the husband and wife.

***Impaired social interaction related to loss of a new born secondary to congenital malformations*:** Allowed her to ventilate her emotions, encouraged her to attend the counselling sessions and group treatment options to decrease isolation and to expand alternative coping mechanism. Involved husband in the therapy and counseling and provided positive strokes and avoided false reassurance.

***Risk for impaired emancipated decision making related to their reproductive health*:** Provided counselling to both mother and father. Advised for genetic counseling. Assured continuous support when needed.

Mrs. A was able to accept the loss gradually and was receptive for genetic counseling. Family support and counseling helped her to assume normal life.

II. Conclusion

Mermaid syndrome is a rare circumstance with a poor prognosis. Antenatal diagnosis is possible through ultrasound. More emphasis ought to be laid on right prenatal analysis and care with a probable termination of being pregnant proposed if detected early. Prevention is feasible and needs to be the aim. Regular antenatal check takes a look at up with optimal maternal blood glucose degree in pre- conceptional length and in the first trimester needs to be maintained to prevent this anomaly.

Once identified, nurses' role is vital in providing psychosocial and emotional support.

In order to promote an environment where the couple understands and develops coping strategies to overcome their loss. The nurse facilitates a space where the couple can express their thoughts and feelings and are free to ask their queries. The nurse should help them to understand and should help them to engage with their routines effectively[16].

References

[1]. Nadereh Taee, Tarhani et.,al., Mermaid Syndrome: A case Report of a rare Congenital Anomaly in Full-Term Neonate with Thumb Deformity, American Journal of Perinatology.,2018 oct 8 (4)

[2]. Duhamel B (1961) From the Mermaid toAnal Imperforation: The syndrome of caudal Regression. Arch Dis Child 36(186):152-155

[3]. Reddy K, Srinivas S, Kumar S, Reddy s, Hariprasad, et al. (2012) Sirenomelia Arare presentation. Journal of neonatological surgery 1 (1):7

[4]. Singh C, Lodha P, Arora D, Prabhu Sharma A, Kaul A(2014) Diagnosis of sirenomelia. Prenat Diagn 26(8): 684-688.

[5]. Garrido-Allepuz C,Haro E, Gonzalez- Lamuno D et al.: A clinical and experimental overview of sirenomelia: insight into the mechanisms of congenital limb malformations. Dis Model Mech. 2011, 4: 289-99

[6]. Duester G(2008) Retinoic acid synthesis and signaling during early organogenesis. Cell 134:921-931

[7]. Garrido-Allepuz C,Haro E, Gonzalez- Lamuno D et al.: A clinical and experimental overview of sirenomelia: insight into the mechanisms of congenital limb malformations. Dis Model Mech. 2011, 4: 289-99

[8]. Al-Haggar M, Yahia S, Abdel-Hadi D, Grill F, Al Kaissi A. Sirenomelia (symelia apus) with Potter's syndrome in connection with gestational diabetes mellitus: a case report and literature review. Afr Health Sci. 2010;10(04):395–399.

[9]. Chan B W, Chan K S, Koide T et al. Maternal diabetes increases the risk of caudal regression caused by retinoic acid. Diabetes. 2002;51(09):2811–2816.

[10]. Taee N, Tarhani F, Safdary A. A case of sirenomelia in newborn of a diabetic mother. Int J Diabetes Metab. 2005;5(01):83–87.

[11]. Stocker Jt, Heifetz SA: Sirenomelia. A morphological study of 33 cases and review of the literature, Perspect Pediatr Pathol.1987, 10: 7-50
[12]. Stevenson RE, Jones KL, Phelan MC, et al.: Vascular steal: the pathogenic mechanism producing sirenomelia and associated defects of the viscera and soft tissues. Pediatrics. 1986, 78:451–57.
[13]. Khan N Ismail F, Werke I, Gongxeka H (2010) Mermaid baby. SA J radiology 14:66-68
[14]. Saxena, R., & Puri, A. (2015). Sirenomelia or mermaid syndrome. *The Indian journal of medical research*, *141*(4), 495.
[15]. Samal, S. K., & Rathod, S. (2015). Sirenomelia: The mermaid syndrome: Report of two cases. *Journal of natural science, biology, and medicine*, *6*(1), 264–266.
[16]. Eldhose.N.J., Councelling Psychology, Departement of Psychology., University of Calicut, 2014., pg.no. 15.

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