

Mermaid Syndrome

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Abstract

The mermaid syndrome is a very rare abnormality with an incidence of 1 in 100,000 births. In this condition a baby is born with both legs joined together giving them a fish like appearance and most of them has a poor prognosis. Many other systemic anomalies and usually a single umbilical artery is seen with this syndrome. The exact etiology of the condition is not clear but there are some factors identified which are said to be associated with Mermaid Syndrome. No appropriate treatment is advised till now for this syndrome but symptomatic management is initiated.

Keywords: mermaid syndrome, sirenomelia, fused legs

Introduction

Mermaid Syndrome

Mermaid syndrome or Sirenomelia, is a rarest of rare congenital developmental disorder characterized by anomalies of the lower spine and the lower limbs. Affected babies are born with partial or complete fusion of the legs which gives them the appearance of mermaid. Mermaid means trunk looks like human and rear looks like a fish ^[1]. In this particular syndrome the upper part of the body is functioning normally but the lower part of the body is fused together leaving a fish like appearance.



Fig 1

Synonyms of mermaid syndrome

1. Sirenomelia
2. Sirenomelia sequence
3. Sirenomelus
4. Sirenomelia syndrome

Causes

The cause of mermaid syndrome is unknown. Several suggested etiologic factors are as follows,

1. Maternal diabetes is so far only disease condition identified to be associated with this syndrome.
2. Toxic substances or teratogens like retinoic acid, cadmium, cyclophosphamide have reported the cause of mermaid syndrome in mice.
3. Family history of Caudal Regression Syndrome
4. "Vascular steal" theory by Stevenson- The common feature is the presence of a single large artery, arising from high end in the abdominal cavity, which diverts nutrients from the caudal end of the embryo distal to the level of its origin. The steal vessel derives from the vitelline artery complex, an early embryonic vascular network that supplies the yolk sac. Arteries below the level of this steal vessel are underdeveloped and tissues dependent upon them for nutrient supply fail to develop, are malformed, or arrest in some incomplete stage.
5. As a part of VACTERL syndrome (vertebral defects, anal atresia, cardiac defects, trachea-esophageal fistula, renal anomalies, limb abnormalities)
6. Women suffering from severe illnesses during pregnancy
7. Mutation of genes from mother or father
8. Physical & mental stress ^[2, 3].

Signs & symptoms

1. Affected infants may have one foot, no feet or both feet, which may be rotated externally.
2. The tailbone is generally absent and the sacrum is also partially or completely absent.
3. Genitourinary abnormalities- Absence or underdevelopment of one or both kidneys, cystic kidney malformation, absent bladder, urethral atresia, imperforate anus
4. Gastrointestinal abnormalities- esophageal atresia, intestinal malformation, Omphalocele
5. Anomalies of the lumbosacral spine and pelvis-lordosis, Spina bifida
6. Facial abnormality- large low set ears, flat nose, hypertelorism (wide set eyes), Prominent upper eyelid fold.
7. Additional abnormalities-absence spleen, absent

- gallbladder, heart defects, lung defects
- Sirenomelia is often fatal during the newborn period [4].

Classification

Stocker and Heifetz classified sirenomelia or Mermaid Syndrome in seven types-

- All thigh and leg bones present.

- Single fibula.
- Absent fibulae.
- Partially fused femurs, fused fibulae.
- Partially fused femurs, absent fibulae.
- Single femur, single tibia.
- Single femur absent tibiae [5].

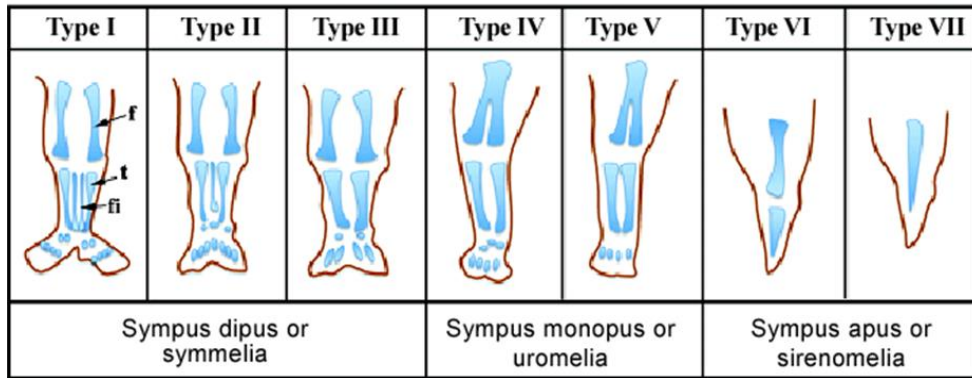


Fig 2: Classification of syrenomelia to Stocker and Heifetz

Diagnostic evaluations

- It's usually diagnosed during second trimester during USG examination.
- After birth, only physical examination is used to make a diagnosis of mermaid syndrome.
- CT scans & MRI scans to assess bones & soft tissues.
- X ray- to assess bone structures.
- USG- to assess any deformity of abdominal organs [6, 8]

Treatment

There is as such no treatment advised for Mermaid Syndrome. Treatment of Mermaid Syndrome during pregnancy: If anomaly is diagnosed during pregnancy, then the healthcare provider may advice on the risks associated with carrying on the pregnancy.

Treatment of Mermaid Syndrome after birth

- Surgical separation of the lower extremity
- In severe cases, the child's lower extremity may have to be amputated
- Colostomy
- Kidney transplant in case of absence of kidneys [9]

Prevention

- There are no reported methods of preventing Mermaid Syndrome.
- Regular antenatal visits to hospital might facilitate early detection of any anomalies in the fetus [10].

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