

## SYRENOMALIA (SYMELIA APUS—?)

### (A Case Report)

by

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The word derives its origin from the word Syreno—meaning a Nymph of the high Seas and Malia with feet like that of a fish: The common name for this deformity is "Mermaid". History and Mythology are galore with stories of Mermaids attacking lone ships in high seas.

One of the earliest to report this uncommon deformity was Bartholin (1654). Ballantyne (1904) reviewed a number of cases. Further cases were collected and reviewed by Kempmeier (1927). Hendry and Kohler (1956), Bearne (1960), Foulkes and McMurray (1954), Bhatt and Venkatramani (1968), Rao, Kumari and Kumari (1960) have contributed some more cases to the existing literature.

Though not rare, this type of congenital anomaly is uncommon. This case appears to be the first documented case report from this hospital during the last five years.

#### Case Report

Mrs. J. aged 30 years, gravida 4, was admitted with labour pains on 26-4-1971 at 10 P.M. The patient was 36 weeks' pregnant. She delivered a live sympodic baby by vertex at 2.45 a.m. The placenta and membranes were delivered spontaneously. The amniotic fluid was not measured but appeared to be lesser than normal. The baby did not show any signs of life.

The patient had an uneventful puerperium. She was discharged after 7 days.

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The patient came from a nearby village. She did not give any history of drug intake or sickness in the present pregnancy. There was no history of previous irradiation. Her two previous deliveries were normal. No person in her family has ever given birth to such a foetus.

The baby weighed about 2.6 Kgs. It showed no signs of life at birth.

#### Postmortem Examination

The crown-rump measurement was 11½". Circumference of the chest 12". Circumference of the thorax 12".

No abnormalities were detectable on the face and pinna, scalp, thorax, or upper limbs. The abdomen appeared normal, the umbilical cord had only one single umbilical artery arising from the aorta. At the lower end there was only one limb attached on the left side of the pelvis, terminating in a small vestige of a nail. External genitalia and anal openings were totally absent. On opening the abdomen, the liver was seen to be occupying much of the cavity. The stomach and the small intestines were normal. The caecum ended in a blind pouch about 4" in length, and it contained inspissated meconium. The abdominal aorta ended blindly at the level of the pelvic inlet. The suprarenal glands, the kidneys, the ureters and the bladder were totally absent. The internal genitalia were also absent.

On opening the thorax, the great vessels were seen to be normal. The heart consisted of single chamber (Cor Biloculare—?) with the aorta arising from the upper end. The lungs were collapsed and failed to float in water.

#### Discussion

Sympodial abnormalities are quite rare though not uncommon. The exact mecha-

nism of formation of these anomalies is yet under much discussion. Following are some of the various views based on certain factors.

1. Changes in quantity of amniotic fluid is regarded as one of the causative factors. A constant feature of symphyal anomalies is oligohydramnios. The scant amniotic fluid probably causes the malrotation acting as a mechanical barrier. Agenesis of kidneys, a common feature with sirenomelia may be responsible for the small quantity of amniotic fluid. No measurement of amniotic fluid was made in this case.

2. A probable injury to the developing foetus in the state when the primitive urogenital and cardiovascular systems are undergoing differentiation may be responsible. This is suggested by the multiple defects in these systems noticed in a sirenomalic foetus.

3. A single umbilical artery is a constant feature. This may interfere with the growing limb 'buds' nutritional status. This was present in this case. Albeit single umbilical arteries, are also noted in normal foetuses.

4. Consanguinity has been held responsible by some.

5. Maternal subfertility has been implicated by others.

6. Elderly primiparity.

7. Failure of development of caudal segments in the differentiating embryo.

8. Experimentally, Wolff, (1936) produced syrenomalic deformities in the chick embryo by irradiating the caudal segments with x-rays.

Potter (1946) reported a case of syrenomelia with characteristic facies, renal agenesis, and pulmonary hypoplasia. A single umbilical artery is a common association. A case with harelip has been reported by Rao.

One often expects a female phenotype in a "Mermaid". On the other hand Potter, showed only 4 females in 26 of her cases, while Kempmeier had 14 females in his 52 cases. The external genitalia are often absent and the internal genitalia are either absent or rudimentary.

During the normal development, the postaxial border and also the little toe become lateral, while the sole faces posteriorly. The knee cap therefore faces anteriorly. Such a rotation fails in a syrenomalic foetus and thus the deformity. Large flipper-like upper limbs have also been noticed. Absence of bony skeleton, presence of fused femora with 5-6 bony appendages have also been noted in some cases. Spina bifida, absence of pelvic bones and sacrum have all been reported earlier. In this case, the femur was single and it articulated with the acetabulum on the left side. The pelvic bone was present only on the left side; the ala of sacrum was absent on the right side.

"Self-differentiation" is very important in the formation of the skeleton. The general form, the limbs, joints and the general configuration of bones are then dependent on extrinsic conditions prevailing in the maternal environment" (Sissons). In a syrenomalic development there may be such a failure of self-differentiation and this may result in the symphyal deformity.

Forster (1861, 1865), classified the (syrenomelia) "Mermaids" into:

1. Symelia Bipus: Fusion of limbs just extending upto the ankles with fusion of both feet simulating that of a fin of a fish. (Mermaid).

2. Symelia Unipes: Presence of a single foot formed by the partial union of both feet. The bony skeleton can be

made out. Multiple toes are also present.

3. Symelia Apus:— No feet are present, both the limbs being imperceptibly merged into one.

#### Summary

A case of syrenomalia—type Symelia Apus (?) (Forster) has been reported, with hemipelvis, spina bifida, absent genitalia, absent renal system, a cor unilocular type of heart, a single umbilical artery and pulmonary hypoplasia.

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