

SIRENOMELIA: SYMPUS DIPUS ("MERMAID")

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Hendry and Kohler (1956) have recently reviewed the condition of sirenomelia, and in view of its rarity it was thought that a further case record would be of value.

Case Record

The mother, a primigravida of 18 years, had a normal pregnancy, she was rhesus negative, and had no antibodies. The infant was delivered as an extended breech on July 6, 1957, four weeks before term, and weighed 3 lb. 14 oz. (1.76 Kg.). Grunting respirations occurred as soon as the child was delivered and these continued for several minutes, while the heart continued to beat for one hour. Immediately after delivery the child cried and the toes were seen to move.

External Examination. At necropsy a well-preserved sirenomelus of uncertain sex, 17½ inches long, was seen (Fig. 1). The upper limbs appeared flattened antero-posteriorly, and the wrist, metacarpo-phalangeal and interphalangeal joints were readily hyperextended. The pelvic girdle was about two-thirds the expected diameter. A small, shallow dimple was present over the top of the coccyx, and another dimple, immediately below it, represented an imperforate anus. External genitalia were not present, nor was there any urethral orifice. The lower limbs were fused in their whole length and were rotated so that the patellae were on the lateral aspect of the leg. Movement at the knee was possible in a forward direction only. The fused feet were inverted, with the soles anterior, the heels posterior, and the great toes lateral. Eight toes were present, but the smallest was in the midline, on the posterior aspect, and was not visible from the front.

The pinnae were misshapen and placed a little lower than normal. They did not contain cartilage.

Internal Examination. The head and neck were normal. In the thorax, a tracheo-oesophageal fistula was present half-way between the larynx and the bifurcation of the trachea. The upper end of the oesophagus ended blindly, and was wider and thicker than the lower part which emerged from the anterior aspect of the trachea and continued normally. The trachea was normal except for the small aperture leading into the oesophagus. The lungs were poorly expanded. The inter-lobar fissures were normal except for the right transverse which was represented by a fibrous band. The thorax was otherwise normal.

In the abdomen, the spleen, pancreas, liver and biliary passages were normal; the only abnormal part of the alimentary canal was the sigmoid colon, which ended blindly in a bulbous swelling lying in the left iliac fossa. The adrenal glands were large, occupying an area equal

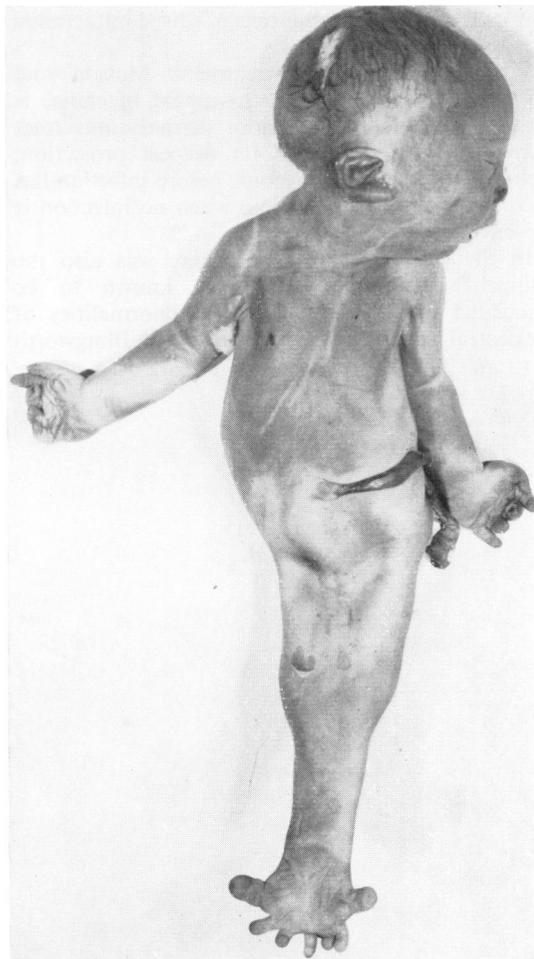


FIG. 1.—Post mortem photograph of sirenomelus.

to that normally occupied by the kidneys. A spherical body, 4 mm. in diameter, situated immediately inferior to the left adrenal gland, was shown histologically to be a rudimentary kidney. No ureters were present but a tubular structure was present on each side below the level of the kidney, attached to a gonad and ending blindly in the skin of the inguinal region (Fig. 2). Histological

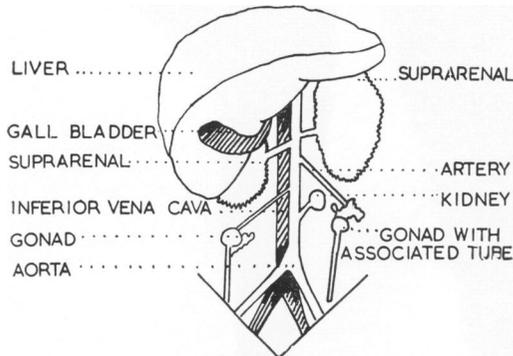


FIG. 2.—Diagram of posterior abdominal viscera.

examination of the gonads showed embryonic seminiferous tubules in the form of solid rods some of which showed early lumen formation, corresponding to the fifth month of development, while cell nuclei examined from several sites showed a female chromatin pattern.

No other pelvic organs were present. The great vessels in the trunk appeared normal, but there was only one artery in the umbilical cord.

MUSCULATURE OF PELVIS AND LOWER LIMBS (Fig. 3). The only internal pelvic muscles present were iliacus and psoas on either side. An unidentified muscle ran between the two greater trochanters. Muscle fibres, arising from both surfaces of the transverse processes of the lower three segments of the sacrum, fused just inferior to the tip of the coccyx, to form a median muscle, whose tendon was inserted into the morphological lateral condyle of the right tibia. The hamstrings were absent. The quadriceps attachments were normal except for an extra branch of the patellar tendon on the left, which ran infero-medially, to be inserted into the tarsus. The deeper fibres of vastus lateralis formed a cruciate arrangement posterior to the knee-joint, and a few of these fibres formed a slender median muscle, whose tendon was attached to the tarsus. The anterior tibial group of muscles (on the posterior aspect of the lower limb) was abnormal and no muscles arose from the anterior aspect of the lower legs. An interosseus muscle arising from the central fibula ran laterally to each tibia.

Radiographic Examination (Fig. 4). The head, neck and upper limbs were normal. There were seven cervical vertebrae, 13 thoracic vertebrae and paired ribs, and six lumbar vertebrae, five sacral segments directed more posteriorly than usual, and a coccyx. In the pelvis, a

median bony ridge represented the fusion of the right and left conjoined rami of the ischium and pubis, thus causing the acetabula to face postero-laterally. In the conjoined lower limbs, two separate femora and two tibiae were present, while a median bone, equal in width to the tibiae, lay posterior to them and was thought to represent a fusion of the fibulae. Three centres of ossification were seen in the tarsus, and eight toes were present.

Discussion

In sirenomelia, the developmental abnormality in the lower limbs appears to be a failure of the medial rotation which normally takes place in foetal life. Consequently the anterior aspect of the limb is directed laterally and the fibulae come to lie medial and posterior to the tibiae. This medial position of the fibulae is a characteristic finding in the condition and in our case there is only a single median bone which from its width and muscle attachments is thought to represent a fusion of the fibulae. The failure of rotation causes the soles of the feet to be directed anteriorly and the great toes to be on the lateral aspect of the feet, and results in the peculiar fish tail shape of the fused feet. There is similar failure of rotation of the thighs so that the greater trochanters are directed posteriorly.

Malformations of the urinary tract are usual in

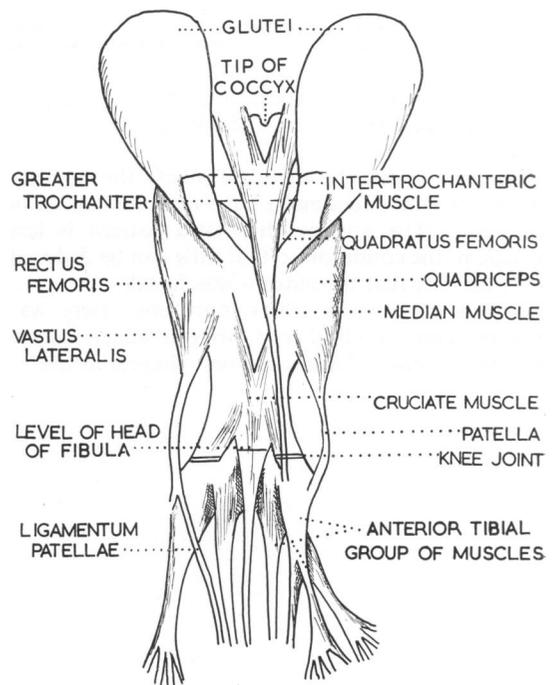


FIG. 3.—Diagram of musculature of pelvis and lower limbs.

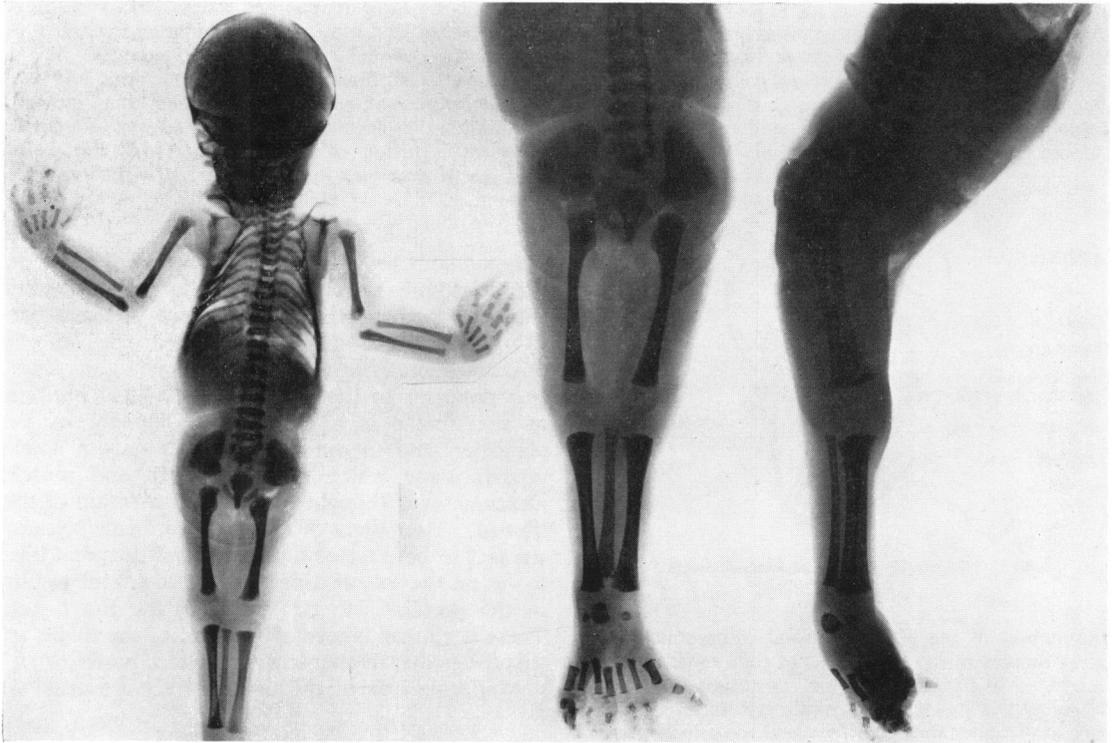


FIG. 4.—Anterior and lateral radiographs of skeleton. The extra vertebrae, the median pelvic bone, the posterior tilt of the sacrum, the posterior fused fibulae, and the fused feet are well shown.

this condition and in this case the only existing part of the urinary system was a single rudimentary left kidney.

Contrary to popular mythical beliefs, the majority of infants with sirenómelia have male gonads as in this case. The nuclear chromatin pattern is less specific to the condition so that little can be deduced from the fact that the pattern was female.

In this case, as in others, only one artery was present in the umbilical cord, while an added feature was the presence of a tracheo-oesophageal fistula.

Summary

A further case of sirenómelia is recorded in view of recent interest in the subject.

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REFERENCE

Hendry, D. W. and Kohler, H. G. (1956). *J. Obstet. Gynaec. Brit. Emp.*, 63, 865.