

**EUGONADOTROPIC AMENORRHOEA
(ROKITANSKY-KUSTER-HAUSER SYNDROME)**

by

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CASE REPORT

Miss. T. S. 24 year old, Hindu, came with the C/o primary amenorrhoea and pain in left inguinal region off and on for 2 months. She had the swelling since 3 months of age. Operation advised during infancy, and childhood was refused by the mother. No history suggestive of pituitary tumour (headache, visual disturbance etc.) No. H/O any endocrine diseases like Cushing's, hypothyroidism or congenital adrenal hyperplasia. No history of tuberculosis or any other major illness like malnutrition, malabsorption, renal and cardiac disease, severe infection or neoplasia.

No H/O taking any drugs.

On general examination she had a normal phenotype of a female, height being 157 cm. and body fat proportionately distributed. Axillary and pubic hair development was normal and breasts were well developed the vital signs were within normal limits (P-84, Bp 110/80) Local examination of perineum revealed, the introitus, clitoris, labia minora majora were symmetrical and normal. There was complete absence of vagina. (as shown in photograph) There was a pouch of $\frac{1}{2}$ cm. in depth in the place of vagina. PR did not reveal the presence of uterus.

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There was left inguinal swelling just above the external ring and was 1 cm. in diameter, non-tender and no impulse on coughing was found. No other congenital malformations were found on examination, On Investigation Hb.-11 gms% Urine—NAD Barboddy+ve, the IVP, Xray Skull and X-ray Chest were normal.

On Laparoscopic examination, the uterus, left ovary and tube were found absent. The right ovary was normal in size and shape and a rudimentary right tube was present. The left inguinal swelling was explored which showed the uterus 2.5 x 2.5 x 1 cm. in dimension with right ovary and tube which were normal in size shape attached, to it, packed compactly in a sac. The sac had no connection to the peritoneal cavity. The sac with its contents was removed.

Histopathology of the uterus showed normal muscles with endometrial cavity. The ovary and tube showed normal structure.

In our case there is a failure in the development of Mullerian ducts and its fusion resulting in the formation of vagina. The uterus was rudimentary and it was found in inguinal region. Deutschman (1923) pointed out the ovaries descend from lumbar region in the 8th wk. of gestation to the true pelvis. In his case he reported the ovaries descended to the inguinal canal dragging the uterus, the tubes along with it as occurred in our case.

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See Figs. on Art Paper VI