

Rudimentary Uteri, the ovaries and vaginal length in MRKH Syndrome

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Background: Mayer-Rokitansky-Kuester-Hauser (MRKH) syndrome is a malformation of the female genital tract due to interrupted embryonic development of the Mullerian (paramesonephric) ducts. The presence of rudimentary uterine buds has been reported but there is uncertainty as to the incidence, size and nature of these structures. Ovarian position and the MRI evaluation of vaginal length in these patients has not been reported.

Aim: To assess the gynaecological tract in patients with MRKH syndrome. The incidence of rudimentary uteri and the level of differentiation, the size and appearances of the ovaries and the length of the distal vagina have been assessed.

Patients and Methods: The MR scans of 40 women under the care of a specialist clinic for Disorders of Sex Development (DSD) with a diagnosis of MRKH syndrome were reviewed. The mean patient age was 20.8 years, range 13-40 with a median of 19 years. The presence of uterine remnants was assessed and uterine volumes were calculated from three orthogonal dimensions of the uteri. Ovarian volume and position were assessed. Vaginal length was measured according to established methodology (1). The presence of differentiation of the uteri into 1, 2 or 3 layers was recorded.

Results: Bilateral rudimentary uteri were found in 35 patients, 1 had a rudimentary uterus on one side and none on the other. Four patients had no uterine structure at all. All uteri were sited laterally in the pelvis, and none were conjoined. A low signal band, likely to represent the fibrous band found surgically, was seen to extend between the rudimentary uteri in a minority of patients. Mean uterine volumes were 4.4 ml on the right and 5.7 ml on the left. Volumes ranged from 0-14.7 ml on the right and 0-31.3 ml on the left.

Sixteen uteruses in 11 patients showed differentiation into more than one layer. There were 7 uteruses showing a 2 layered appearance and 9 showing 3 layers. One patient had a uterus with 3 layer differentiation and intraluminal blood on one side and a streak uterus on the other.

Bilateral ovaries were present in all patients. The mean ovarian volume was 9.0 ml on the right and 8.3 ml on the left. Seventeen patients had ectopically sited ovaries (one or both). These were most commonly found either very antero- laterally, or high in the pelvis at the level of the pelvic brim.

There was a constant relationship between the ovaries and the uteri- the uteri were always found immediately related to the ovaries even when the ovaries are ectopic.

A perineal dimple only was found in 12 patients. In the other 28 patients, vaginal length was ≤ 1 cm in 6, > 1 cm ≤ 2 cm in 9, >2 cm ≤ 3 cm in 10. In 3 patients the vaginal length was 3.8 cm, 4.3 cm and 6.5 cm. The latter is within the normal range (2).

Discussion: Rudimentary uteri are common in patients with MRKH and these can be relatively large (up to around 30 ml). In one patient the uterus was distended with blood from functioning endometrium and this could be an unsuspected cause of pain. Ovaries are commonly ectopically sited, but the uteri remain closely related to them. This is of interest embryologically as they arise from different tissue. The vagina can be measured from MR scans thus obviating the need for vaginal examination in some patients (e.g. young or sexually inactive patients). Length of vagina can be used as an indicator for the appropriate use of vaginal dilation therapy(3).

Reference:

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