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RESEARCH ARTICLE

PERSISTENT MULLERIAN DUCT SYNDROME (HERNIA UTERI INGUINALE).

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Abstract

Ambigous genitalia comprises of male pseudohermaphrodite, female pseudohermaphroditism,mixed gonadal dysgenesis, and true hermaphrodite. We are dealing with male pseudohermaphrodite in this article, Transeverse testicular ectopia is a rare disorder which arises from a lack of anti Mullerian hormone(AMH or Mullerian Inhibiting substance-MIS.)MIS acts on the Mullerian ducts, resulting in the presence of Mullerian structures in a normally virilized XY male. PMDS is due to either a mutation in the AMH gene or from a defect in the AMH type II receptor. These mutations display an autosomal recessive mode of inheritance.
Risk of testicular malignancy is the same as that for an intra abdominal testis. Testicular tumours such as seminoma , non seminomatous germ cell tumours, testicular carcinoma and clear cell adenocarcinoma have all been reported and can cause obstructive uropathy.

Introduction:-

We had seen 5 cases of hernia uteri inguinale otherwise called transverse testicular ectopia in **ICH-Madras Medical College** over a span of ten years from 2007-2017.

Clinical features of bilateral undescended testes seen in these children. Inguinal hernia was seen usually in left side . Ultrasonogram showed gonads in inguinal region in 2 cases. KARYOTYPING done in these children proven to be XY . AntiMullerian hormone level were less .Testosterone levels were below normal. GONADAL biopsy showed atrophied seminiferous tubules with hyalinization.Through inguinal incision inguinal canal opened and gonads attached with mullerian remnant was divided preserving vas and vessels,and both testes brought to subdartos pouch and orchidopexy done.

It is a autosomal recessive disorder which arises that arises from a lack of anti Mullerian hormone(AMH or Mullerian Inhibiting substance-MIS) action on the Mullerian ducts, resulting in the presence of Mullerian structures in a normally virilized XY male. PMDS is due to either a mutation in the AMH gene or from a defect in the AMH type II receptor. Risk of testicular malignancy is the same as that for an intra abdominal testis. Testicular tumours such as seminoma , non seminomatous germ cell tumours, testicular carcinoma and clear cell adenocarcinoma have all been reported .

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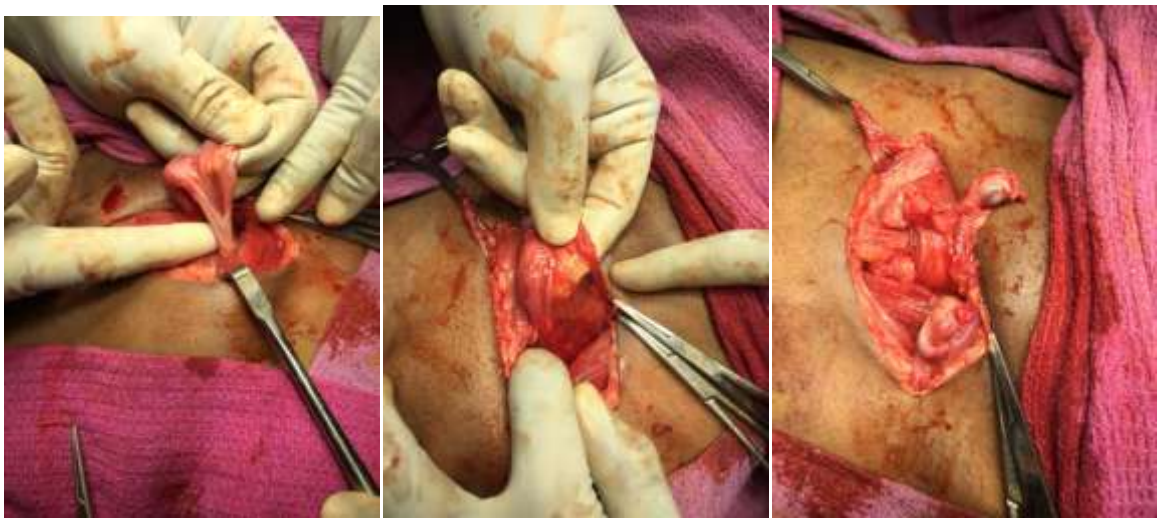

Conclusion:-

To avoid devascularistion of vas ,myometrium is left intact along the length of vas.Division of mullerian remnant prevents cyclical hematuria in future. In all cases of cryptorchidism with inguinal hernia ,do magnetic resonance imaging to look for transverse testicular ectopia and measure anti-mullerian hormone levels. These patients are usually azoospermic and have to bekept. under follow up and malignancy changes of testes should be noted after puberty.

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