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A Case of Hydrometrocolpos due to Persistent Urogenital Sinus: Mckusick - Kaufman's Syndrome

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Hydrometrocolpos results from vaginal obstruction, either from a simple imperforate hymen or from a high vaginal atresia.

One serious form of vaginal obstruction is that associated with the urogenital sinus. In this anomaly, examination of the perineum reveals two openings: the normal anus and a single, anterior common channel opening into the vestibule, the urogenital sinus. This may be mistaken for a normal vagina except for the fact that the urethral orifice is not in its normal location. This is a rare anomaly.

Case report: A 2 months old baby was admitted on 20-1-99 at 9.40 pm with H/O sudden distension of abdomen and difficulty in passing urine.

O/E: All the basic parameters were within normal limits. Abdominal examination revealed a firm mass occupying the rt iliac fossa, rt lumbar, umbilical, hypogastric and lt iliac regions. Mobile. Splenomegaly +.

O/E: The external genitalia revealed a blind ending vagina. Urethral orifice abnormally located. A provisional diagnosis of hydrometrocolpos with urogenital sinus was made.

U.S.G. confirmed the diagnosis.

Management: Romenosky, Rotfensperger operation (abdomino-vagino-perineal pullthrough).

Baby was put in the lithotomy position. Abdomen was opened by infraumbilical midline incision extended above the umbilicus. Distended vagina was extending above the umbilicus pushing the bladder anteriorly. Vagina (intra abdominal) opened between stay sutures. Purulent collection about 500ml drained.

Finger introduced through the intra-abdominal vaginal opening and vaginal wall pushed into the perineum. Vaginal opening created over the pushing finger. Inverted 'U' shaped skin flap developed in perineum and stitched to the posterior vaginal wall. Rubber tube kept in the neovagina.

Abdominal vaginal opening closed. Peritoneal wash given. Drainage tube kept.

Abdomen closed. Drainage tube removed on the 2nd Post op day.

Rubber tube removed from the neovagina on the 4th post op day. Baby was discharged on29.1.99. When the baby came for the Ist followup after 10 days on 8.2.99, the vaginal opening was quite normal. Baby was followed up for 6 months, and was doing well. Afterwards the baby did not turn up for follow up.

The association of hydrometrocolpos with polydactyly and congenital heart disease is thought to be an autosomal recessive syndrome. Major urinary tract anomalies, including ectopic ureterocele, right renal agenesis and bilateral renal dysplasia, have also been reported.

In view of the large number of associated anomalies in these children, their entire urinary tract should be examined. It is quite likely that a study of the family pedigree would reveal familial genetic traits.

Eventhough incision of the vagina through the urogenital sinus and catheter drainage after identification of the distended vagina at laparotomy is suggested as a

treatment when this technique was used, urine refluxes into the distended, poorly drained vagina and causes persistent sepsis. This complication as well as recurrent stenosis of the incision site, can be avoided by the use of an abdomino-perineal-vaginal pullthrough operation, described above. This created a skin and mucosa lined opening for the vagina on the perineum. This operation leaves the entire urogenital sinus as the urethra.

These infants should have a speculum examination under anaesthesia at intervals after the operation to observe healing and to prevent stricture at the anastomosis.