IMPERFORATE VAGINA IN THE NEWBORN*

NEONATAL HYDROCOLPOS

BY

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The purpose of the present communication is to discuss hydrocolpos and hydrometrocolpos, a not uncommon complication of imperforate vagina in the newborn. We do not propose to discuss the relatively common problem of vaginal occlusion due to labial adhesions (Fig. 1), which can be easily separated with little or no bleeding.

Hydrocolpos in the newborn female due to imperforate hymen or to occlusion by a membrane in the lower third of the vagina is not excessively rare. At or shortly after birth the infant presents with lower abdominal swelling and a bulging membrane is seen at the vaginal orifice (Fig. 2). This bulging membrane looks like a cystocele and when gently reduced the urethral orifice can be seen anteriorly. Occasionally there may be a tiny opening in the occluding membrane through which a white mucoid material can be extruded. The anomaly has been fully described by Mahoney and Chamberlain (1940), by Gross (1953), by Caffey (1956), and by many others and for the purposes of the present communication a brief description of their findings will be sufficient.

In this condition the vagina is invariably greatly distended—hydrocolpos—and on occasions the accumulated fluid can stretch the cervical canal and the body of the uterus as well—hydrometrocolpos—though we have seen the uterine cavity distended only on one occasion. The ballooned genital tract arises from the pelvic floor into the abdominal cavity. The cyst is filled with clear or mucoid fluid which tends to become purulent from bacterial invasion, giving rise to pyocolpos. The uterine tubes have not been involved in any of the reported cases. The distended genital tract may cause urinary obstruction and the enlargement of the bladder may hinder palpation of the vaginal cyst. Megaloureter and hydronephrosis may follow and pyuria will be a further complication. Posterior pressure may cause rectal obstruction. There may be respiratory embarrassment through the upward displacement of the abdominal contents. On digital rectal examination the pelvis is filled by a mass projecting against the hollow of the sacrum. On inspection of the vagina a bulging membrane is seen at or above the hymen. In such patients a needle is inserted through the membrane and fluid is aspirated to confirm the diagnosis. Radiopaque fluid may be instilled through the membrane and fluid is aspirated to confirm the diagnosis. Radiopaque fluid may be instilled to outline the obstructed vagina (Fig. 3). Having made the diagnosis the occluding membrane is slit with a knife and the cyst contents are evacuated.

Less commonly the hydrocolpos is due to atresia of the lower vagina (White and Dennison, 1958) and

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in our limited experience of five cases the diagnosis was finally confirmed only at laparotomy. One of us (W.M.D.) first encountered this condition some 10 years ago and the account of his misdemeanours may serve as an awful warning to any junior colleague. A very ill baby, 1 day old, was admitted suffering from peritonitis and intestinal obstruction. At laparotomy dense adhesions were found and a large supravaginal (?) urachal cyst was demonstrated. This was freed, the ureters dissected off and the cyst excised in toto. When the specimen was examined later, it became evident that the operator had performed what is probably the earliest panhysterectomy on record! The so-called urachal cyst consisted of a grossly distended vagina on top of which was a tiny uterus complete with tubes and ovaries. The patient did not survive surgical intervention.

Figs. 4 and 5 show in a schematic fashion the essential anatomical differences between vaginal occlusion by a membrane, a simple well-known problem and the relatively undocumented hydrocolpos due to vaginal atresia. In spite of the anatomical differences the clinical features and the
complications are similar when the hydrocolpos is due to atresia of the vagina. During the past few years we have been particularly unlucky to encounter five infants with atresia of the lower vagina. In all these cases there was a solid block of tissues (at least 2 cm.) separating the distended vagina from the perineum and there was no bulging membrane present to aid diagnosis. In spite of this we have confidently diagnosed hydrocolpos due to vaginal atresia in all these infants, four babies and one girl aged 7 months. Unfortunately only one of our five patients is alive and well and she has lost both her vagina and her uterus.

In the newborn babies dense adhesions make exploration of the abdomen difficult. The cysts vary from 10 to 15 cm. in diameter and in two of the four babies the 'cyst' contents were grossly infected (pyocolpos). In only one case was the uterus distended (hydrometrocolpos). On three occasions the cyst was evacuated through the abdomen and no attempt was made to drain the vagina via the perineum. In the uninfected case it was theoretically possible to evacuate the cyst through the abdomen and after dissecting down towards the lower pole of the cyst to pass a probe through the inferior pole of the cyst until it presented behind the urethra in the region of the posterior fornix. A perineal incision over the probe allows drainage of the distended vagina through an indwelling Malecot catheter. It was difficult to visualize the next stage, as it was assumed that the resulting fibrosis following repeated dilatation would make a later McIndoe and Banister (1938) operation almost impossible. In anticipation it was thought that it might be possible to maintain the passage by using a split-skin graft over a mould until the patient was old enough for construction of an artificial vagina. Having learned from our bitter experience, Mr. John Grant, of our Staff, recently dealt with a non-infected hydrocolpos in an older infant. Before evacuating the cyst he carefully dissected down into the pelvis only to find that suddenly an intact cyst was delivered into the abdomen (Fig. 6). The only reasonable solution to the problem then was to separate the tubes and the ovaries, to remove the vaginal cyst and the uterus and to leave the ovaries behind. This infant made an uneventful recovery and remains well.

From the aforesaid it will be evident that we in Glasgow, as yet, do not know how to deal successfully with hydrocolpos or hydrometrocolpos due to vaginal atresia. On the other hand, to the best of our knowledge, there is no evidence of any more encouraging results from other centres.

One could therefore legitimately ask: Why is the anomaly so rare? Haematocolpos at or shortly after puberty is well known. Due to an imperforate hymen the uterine discharge collects within the vagina and the girl presents with abdominal or pelvic pain. There is tenderness on suprapubic pressure and the ballooned vagina can be felt on rectal examination. This condition should be suspected in a girl with pubic hair and enlargement of the breasts who has not yet menstruated. Why do these girls not present with hydrocolpos in the neonatal period? Obstruction of the vagina alone does not normally give rise to symptoms until puberty. One must therefore postulate that apart from the vaginal obstruction, which provides the necessary anatomical conditions, the real cause lies in the over-abundant secretory activity of the neonatal uterine or cervical glands.

Fig. 6.—Intact vaginal 'cyst' and uterus.

In order to find some support in favour of this postulate we have been reinvestigating during the past four years certain aspects of the development of the vagina and also some features of the physiology of the neonatal reproductive organs. Our findings and conclusions, together with the discussion of the relevant literature, will be presented elsewhere and for the time being we shall confine ourselves to the following brief statements.

Contrary to the most generally accepted view in
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the Anglo-American textbooks, according to which only the lower fifth of the vagina is formed by the fused 'sino-vaginal bulbs' growing out from the dorsal wall of the urogenital sinus, we believe with Vilas (1932), Politzer (1955) and Bulmer (1957) that the boundary between the sinus and Mullerian derivatives of the human vagina lies either near to the portio vaginalis or in the cervical canal. We can also confirm Politzer's (1955) observations on the surprisingly great variability of the grade of development of the vagina in foetuses of similar ages and we believe that sometimes the neonatal vagina and uterus can be too immature histologically to be able to respond to hormonal stimuli in the normal fashion.

It is accepted that in general the uterus of the newborn shows a considerable degree of precocious maturity and there is a marked hyperplasia of the lining epithelium and of the glands (Rosenthal and Hellman, 1952). In fact these glands produce a copious mucus secretion, presumably, under the impact of oestrogens and progesterone derived both from the placenta (Brown, 1959) and from the foetal ovaries (Ober and Bernstein, 1955). While there is a tacit assumption that this mucus secretion is largely the product of the glands of the body of the uterus, our own observations revealed a surprisingly greater maturity of the glands of the cervical canal and we believe that these glands are the main, if not the exclusive source of the mucus discharge. With regard to the hormonal control of the secretion of the cervical glands of the uterus there is accumulating evidence to indicate the cardinal role played by oestrogens (Moricard, 1936; Watson, 1939). Later it is shown (Aberbanel, 1946; Zondek and Rozin, 1954; Macdonald and Sharman, 1959; Stern, 1960) that administration of progesterone inhibits cervical mucus secretion.

Most authorities remark on the great individual variations that exist in the urinary output of oestrogens during pregnancy and we believe that the non-appearance of hydrocolpos in some cases of imperforate vagina of the newborn may be due to the low output of oestrogens or in some instances to the blocking effect of increased discharge of progesterone or progesterone-like substances from the placenta or more likely from the foetal adrenal cortex.

While the above two hypotheses could account for the dormancy in an odd case of imperforate vagina, it would stretch the imagination too far to expect them to account for the large number of cases observed in the adult by Bryan, Nigro and Counseller (1949), by McIndoe (1959) and by Simmons (1959). Clearly we must look for some other explanation. In this respect it seems to us highly significant that in the majority of the adult cases of congenital absence of the vagina or of vaginal atresia, palpation or laparotomy reveals ovarian agenesis or some other malformation of the internal reproductive organs. Thus, while in the cases occurring among newborn babies and small infants the abnormality is strictly and exclusively confined to the vagina, a derivative of the urogenital sinus, in the adult cases both urogenital sinus and Mullerian structures are involved and create an anatomical situation with entirely different physiological behaviour and clinical manifestations. Consequently we believe that the two groups should be treated as separate clinical entities which can be usefully compared but should not be confused with each other.

Our main objective in putting forward these somewhat provocative ideas is two-fold: partly we seek advice from our learned audience concerning the acceptability of these views and partly we hope that by reaching some agreement we may gain a closer understanding of the aetiology and also obtain a necessary theoretical basis for the correct treatment of neonatal hydrocolpos and hydrometrocolpos due to vaginal atresia.

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