

**Fœtus showing a rare Mal-development of the Cloaca,  
Procto-dael Depression, Mullerian Ducts and  
other Structures.**

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THE fœtus here described was sent to the Pathological Museum of the University by Dr. Tracy Lydall, to whom I am indebted, not only for the specimen but also for the following account of the labour, which I reproduce in his own words.

"Mrs. S., æt. 32, who had already had five children, engaged me to attend her in her sixth confinement, which she reckoned would take place on or about September 9th, 1903. She is a tall, well-made woman, who had always had normal labours and strong healthy babies. The course of her pregnancy on this sixth occasion resembled very much those which had preceded it, except perhaps that she was rather larger than in any of the others; she felt the child's movements quite plainly from mid-term. None of her previous children had manifested congenital abnormalities of any kind, nor was there history of such on either the maternal or the paternal side. On June 25th (Mrs. S. being at this date nearly at the end of the seventh month of her pregnancy) her husband was severely injured at his work by the explosion of a heated steel tube, which burned him extensively just above the pubes and in the right groin. He was taken to hospital, where he was detained for a period of two weeks. On his return home it was necessary for his wife to dress his wound every day. Mrs. S. was very much shocked on seeing her husband's injuries. She informed me that there was "a hole in the wall of the bowels you could put your hand into," and she at once made up her mind that the child which she was bearing would be marked in some way in consequence. I was sent for urgently about 1 p.m. on August 28th, and found my patient in strong labour. The os was fully dilated and a bag of waters presented, through which could be felt the vertex in the L.O.A. position. I ruptured the membranes. The head was gradually born without assistance. The pains continued, but no advance occurred, and the child's face became deeply cyanosed. In spite of strong pressure on the abdomen above, combined with traction on the head below, no progress was made. I then with some difficulty passed thick tapes under the axillæ of the child and made strong traction on them. The shoulders and arms were soon delivered, but still the rest of the body did not pass. Traction on the arms was resorted to, and whilst pulling thus a sudden sense of giving way was experienced and the rest of the child was born immediately. It was at once apparent that a large umbilical hernia had caused the obstruction, it being wedged at the pelvic brim. This, however, ruptured during traction on the arms, and the trunk was able to enter the pelvis and allow the fœtus to be born. The child cried lustily, and the cord which arose from the upper part of the hernial covering was tied and divided. The placenta came away spontaneously with the next pain. The mother, in spite of the hard time she had passed through, made an admirable

recovery. The hernia, the coverings of which were torn through, as above described, seemed to consist mainly of a distended transverse colon with some coils of small intestine. It was evident that nothing could be done for the child, so it was wrapped in wool and given at regular intervals diluted milk containing 5 minims of brandy to the ounce. In addition to the hernia the child presented many abnormalities, amongst others club-feet, spina bifida and imperforate anus. The most remarkable feature, however, was the aspect of the external genitals. These resembled the female type in showing labia majora, but hanging from between them by a very thin pedicle was a small translucent bladder, about one inch in length and half an inch in breadth at its widest part; it tapered to a point at each end. This bladder did not seem to alter at all during the time the child lived, neither shrivelling up nor becoming more distended. As far as could be made out there were neither vaginal or urethral orifices, and the child was not observed to pass water during its life. The hernial contents protruded more and more as time went on, the colon becoming distinctly more distended and exuding a thin sanguineous liquid which smelt fæcal. The child lived about 48 hours and cried incessantly during that time. No nourishment could be retained, and in view of its condition treatment was quite hopeless."

The fœtus presented a series of congenital abnormalities, many of which were at once evident upon a naked-eye examination of the body. The feet were in a state of double talipes varus. A well-marked median dimple existed over the lower part of the lumbar vertebræ. The anterior abdominal wall showed a large congenital deficiency in the region of the umbilicus. The diameter of this opening was 3 inches. The umbilical cord was attached to the upper margin of the opening, and through the aperture abdominal viscera were protruding. The most important content of this hernial protrusion was a large, thick-walled, fluctuating sac, occupying the lower half of the projecting mass. Transverse colon was recognised above, and between this and the fluctuating sac one or two coils of small intestine peeped out. Each of the projecting viscera was of a bluish-black colour and covered by inflammatory lymph. There was no hernial sac present when the fœtus arrived at the museum. On examining the perineum it was found that there was no anal, vaginal, or urethral orifice present. In the position of the external genitals were two low loose folds of skin, one on each side of the middle line, and united behind resembling a scrotum much more than labia majora. The folds were afterwards found to contain fat. Labia minora, hymen and clitoris were to all appearance absent. In the middle line between these imperfectly formed labia, and rather nearer their anterior than their posterior end, was a well-marked perineal dimple. The deepest part of the dimple was covered by smooth, shining skin, and from it arose the peduncle of a pear-shaped bladder (see photograph No. 1). This bladder or vesicle was thin walled, of a purplish-brown colour, and possessed a moist and glistening surface. It was translucent and moderately tense with

fluid contents. Its length from its attachment to its most dependent part was 2 inches, and the breadth of its broadest part was  $\frac{3}{4}$  of an inch. Overlying the sacrum and coccyx was a large pad of fat of a triangular shape, the apex of the triangle being towards the perineal depression. There was no evidence of any median furrow representing a gluteal cleft. On dissection, a very interesting condition of the pelvic viscera was revealed. The urinary bladder was identified and lay in the lower part of the hypogastric region. There was no evidence of a patent urachus. The bladder was opened on its anterior surface, and clear urine at once began to escape. At the same time the thick walled sac which projected into the umbilical hernia, and which passed down behind the urinary bladder, began to diminish in size. This sac was then opened and was found to be full of clear, straw-coloured fluid, identical with and continuous with the contents of the normal urinary bladder. The urinary bladder contained about two ounces of urine, whereas the large sac behind it contained six ounces. The communication between the two urinary reservoirs admitted a No. 8 urethral bougie, and was situated immediately below and internal to the opening of the right ureter. The left ureter opened into the bladder close to the ureteral opening of the right side. The bladder wall was considerably hypertrophied, being 5 millimetres in thickness. Both ureters were dilated and hypertrophied, their lumen admitted a No. 14 urethral bougie. The hypertrophy was due to an increase of the muscular tissue of the ureter. The wall of the ureter was 4 millimetres in thickness. The kidney on each side was normal; they showed neither dilatation of the kidney-pelvis nor any change in the histological structure of the kidney substance. The urethral orifice of the bladder was situated about half-an-inch in front of the orifices of the ureters.

On passing a No. 6 urethral bougie along the urethra from the bladder, it was found to pass freely for a distance of  $1\frac{1}{2}$  inches beyond which point it would not advance. In this position the tip of the bougie was found to lie immediately behind the bottom of the perineal dimple. It was then ascertained that the fluid contents of the translucent cloacal sac, which measured about one ounce, could be squeezed out along the urethra into the bladder. Microscopical sections were made of the wall of the perineal sac and of its pedicle up to the point where it was attached. It was very disappointing to find that the tissue of the sac wall and peduncle was undergoing a degenerative change very much allied to coagulative necrosis, due no doubt to its increasingly deficient blood supply. The photomicrographs of this sac are not satisfactory for this reason, and they reproduce very inadequately the appearances revealed by a careful perusal of the microscopical sections. I am quite satisfied that this

sac was lined both inside and outside by an epithelial layer. The outer surface is lined by a stratified layer of cells of considerable thickness. The outlines of the cells are very indistinct in many places. Low papillæ are seen in certain positions. Within this layer and immediately beneath it are glandular spaces lined by a single layer of epithelial cells. Many of these acini open upon the surface (see photo-micrograph Fig. 4). The inner surface of the sac is lined by very irregular cells sometimes in one layer, generally in several layers (see photo-micrograph Fig. 5). The tissue lying between the two epithelial layers consists of a loose fibrous stroma, containing a few bundles of non-striped muscle. The pedicle of the sac consists of fibro-muscular tissue lined on the outside and on the inside by epithelial layers similar in all respects to that just described. The photo-micrograph Fig. 6 is taken across the central point of the pedicle and represents the central canal, lined by epithelium and surrounded by a layer of fibrous tissue, and outside which is fibro-muscular tissue.

On opening up the large sac which lay posterior to and communicated with the bladder, it was found to be bi-locular, the two loculi communicating by a canal admitting the index finger. The upper loculus which projected through the umbilical deficiency was oval in shape, being  $2\frac{1}{4}$  inches long and 2 inches wide. The lower loculus, slightly the larger, was  $2\frac{1}{2}$  inches in a vertical diameter. The loculi are well seen in photographs No. 2. The upper loculus, as I will show presently, is the right half of a double uterus, and the lower loculus is the dilated vagina of the same side.

On examining the upper loculus it was found that its internal surface was studded with low nodular elevations most numerous towards its summit. Passing along the entire length of this loculus are two well-marked raphe, one on the posterior and one on the anterior wall, the posterior raphe is rather nearer the middle line of the body than the anterior. The posterior raphe is seen in the photograph. Both raphe extend from the small aperture into which has been inserted a small rod, downwards as far as the oblique ridge immediately below the inter-locular constriction. From each longitudinal raphe oblique furrows pass outwards similar in all respects to those in the normal cervical canal of the uterus. Microscopical sections of the wall of this upper loculus show the following characters:—

The internal lining membrane consists of a single layer of spheroidal or flattened cells; this, however, is wanting in certain situations. This inner surface presents eminences consisting of loose areolar tissue and depressions, both lined by the same layer of cells (see photo-micrograph No. 7). No racemose glands have been seen. Lying subjacent to the epithelial layer is a loose layer of

areolar tissue containing lymph spaces and thin-walled blood-vessels. The nodular elevations are largely composed of this tissue. Sub-jacent to this loose areolar tissue is the non-striped muscular tissue of the sac, arranged in transverse, oblique and longitudinal lamellæ. External to the muscular coat is a layer of loose areolar tissue whose meshes are everywhere engorged with blood, a circumstance which accounts for the bluish black appearance of this exposed part of the hernial content.

The average thickness of the wall of this upper loculus was 5 millimetres. The lower loculus was wholly inside the abdomen and situated in the hypogastric region. In contrast to the upper loculus it was of a pinkish colour. Anteriorly it was covered by peritoneum, posteriorly it was related to the promontory of the sacrum and overlooked the brim of the pelvis; this aspect was devoid of a peritoneal covering. The internal lining membrane of this loculus was smooth, and there were no evidences of anterior or posterior columnæ rugarum. The most dependent part of this lower loculus was just within the upper strait of the pelvis immediately behind the trigone of the bladder. At this point also it comes into close relation with the vagina of the opposite side, from which it was separated by a thin, semi-transparent membrane. Sections were made of this membrane, and it was found to consist of a thin layer of fibro-areolar tissue lined on each side by stratified squamous epithelium (see photo-micrograph No. 8). The wall of the lower loculus, except at this inter-vaginal point, had an average thickness of 5 millimetres. It consisted of the following layers from within outwards, an irregular layer of stratified squamous epithelium, non-striped muscle in transverse and longitudinal lamellæ, a loose layer of areolar tissue, and outside all a single layer of single squamous epithelium representing peritoneum. Passing backwards and outwards from the apex of the upper loculus is a normal Fallopian tube. This represented the Fallopian tube of the right Müllerian duct. It was about 2 inches long and was bound firmly down to the posterior surface of the upper loculus. Close to the free end of this tube and adherent to the posterior surface of the upper loculus was a small, elongated, flattened body, about  $1\frac{1}{2}$  centimetres in length, which on section proved to be a normal ovary. A probe could be passed into the Fallopian tube from the upper loculus. The opening of this tube is seen in photograph No. 2, and the microscopical appearance of the tube is well seen in the photo-micrograph No. 9.

Immediately to the left of the lower loculus and on a slightly more posterior plane is the normal development of the left Müllerian duct, namely a vagina, uterus and single Fallopian tube. Behind the free end of this Fallopian tube is a normal ovary, 1 centimetre in length. The Fallopian tube on this side is tortuous and has a length

of  $\frac{3}{4}$  of an inch. There is no proper fundus to this left-sided uterus, which has a length of  $1\frac{1}{4}$  inches and a breadth of  $\frac{1}{2}$  inch. The right aspect of this small uterus is adhering to the left aspect of the right vagina. Its position and relations are well seen in the photograph. The cervix of this uterus projects with a typical os into a small vagina. This vagina was a small cavity lying to the left of the most dependent part of the lower loculus or vagina of the right side. On being opened it was found to contain about two drachms of white, creamy substance; its vertical length was 1 inch and its breadth about  $\frac{1}{2}$  inch. Its most dependent part was adjacent to the left lateral part of the trigone of the bladder where it ended quite blindly. It was separated from the right vagina by a thin membrane. Into the upper part of this left vagina projected the cervix of this left unicornute uterus, producing fornices quite similar to those in the bicornute uterus. On splitting the cervix of this left uterus, it was quite easy to recognise, with a lens, an anterior and a posterior median raphe, with the typical arbor-vitæ appearance common to the normal cervix. There was no evidence of anterior and posterior columnæ rugarum in the left vagina. The histological structure of the left vagina was similar to that on the right side, the thickness of its wall being 1.5 millimetres. The descending colon ended in a conical blind tube opposite the brim of the pelvis, immediately behind and to the left of the right vagina.

The true pelvis was almost entirely filled up with fat and areolar tissue, the only canal traversing it to any extent being the urethra. From the most dependent parts of the two vaginae to the perineal dimple there was a distance of  $1\frac{1}{2}$  inches entirely occupied by fat and areolar tissue. The true pelvis was conical in type, its outlet being markedly contracted. A dissection was made over the lumbar dimple, but no spina-bifida was found. The inguinal canals were empty.

*Summary.* To briefly summarise the malformations of the pelvic floor and pelvic contents we found the following:—Complete absence of urethral, vaginal and anal orifices; a single, perineal depression, which was not subdivided into urogenital and anal portions. There was, therefore, no perineal body. Labia majora were present and united behind the perineal depression in a scrotum-like posterior commissure. There was an entire absence of labia minora, clitoris and hymen. From the bottom of the perineal dimple a stalked vesicle arose, whose internal cavity was continuous with the urethra. The urinary bladder was normal except for the vesico-vaginal fistula. The urethra was normal except for the marked narrowing before opening into the perineal vesicle. A complete double uterus and vagina were present. The right vagina communicated with the urinary bladder just internal to the right ureteral opening. The right vagina and uterus were distended with urine. The right uterus

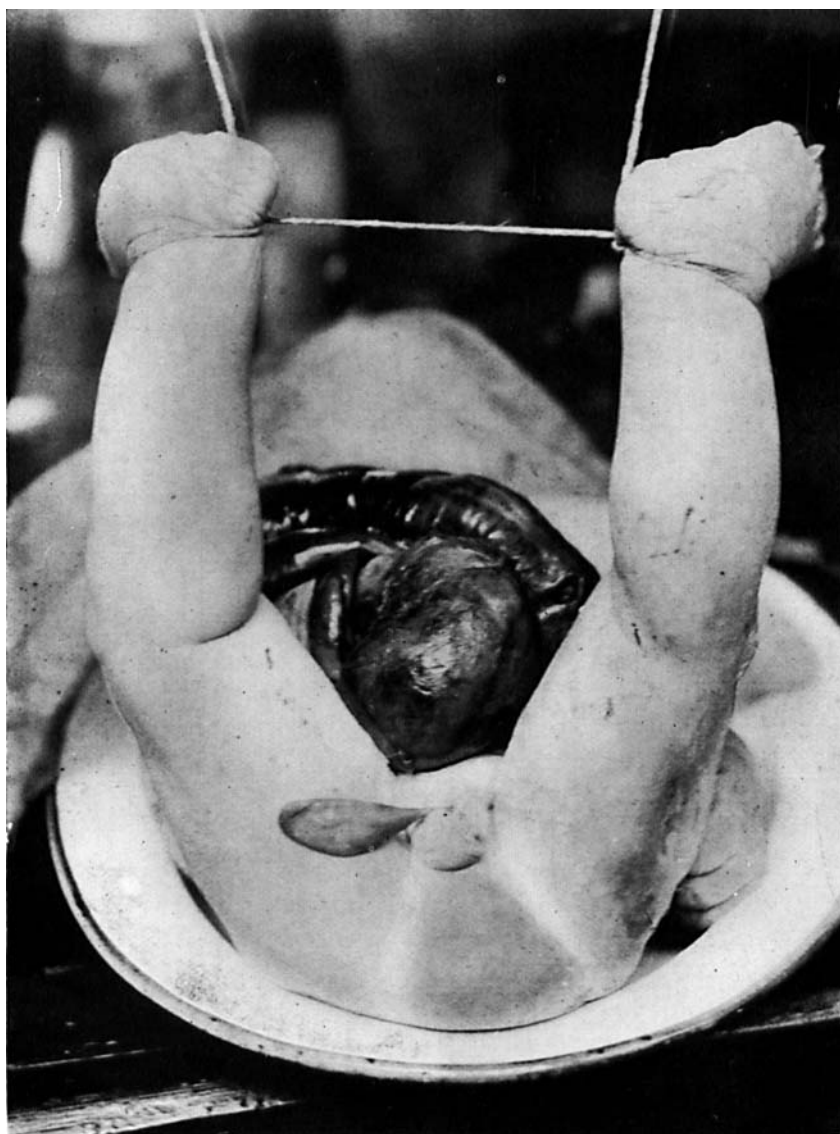


FIG. 1.—Fœtus in lithotomy position, showing perineal vesicle, scrotum-like posterior commissure; umbilical hernia, with upper loculus projecting at lower part; colon above; double talipes, triangular, sacro-coccygeal pad; no gluteal cleft.

Post-median  
raphe.

Opening of  
Fallopian Tube  
(right).

Right Uterus.

Window in  
Bladder.  
Shewing rods in  
Vesico-Vaginal  
Fistula and  
also in Urethra.

FIG. 2.—Specimen shows the posterior median raphe in right uterine cavity, passing from opening of Fallopian tube to ridge between vaginal locus and uterine locus.



Right Uterus  
with Post-  
median raphe.

Right Vagina.

Rod passing  
through Vesico-  
vaginal fistula.

Left Fallopian  
Tube.

Left Uterus with  
os uteri ext.

Left Vagina.

Left Ureter.

Right Ureter.

Bladder.

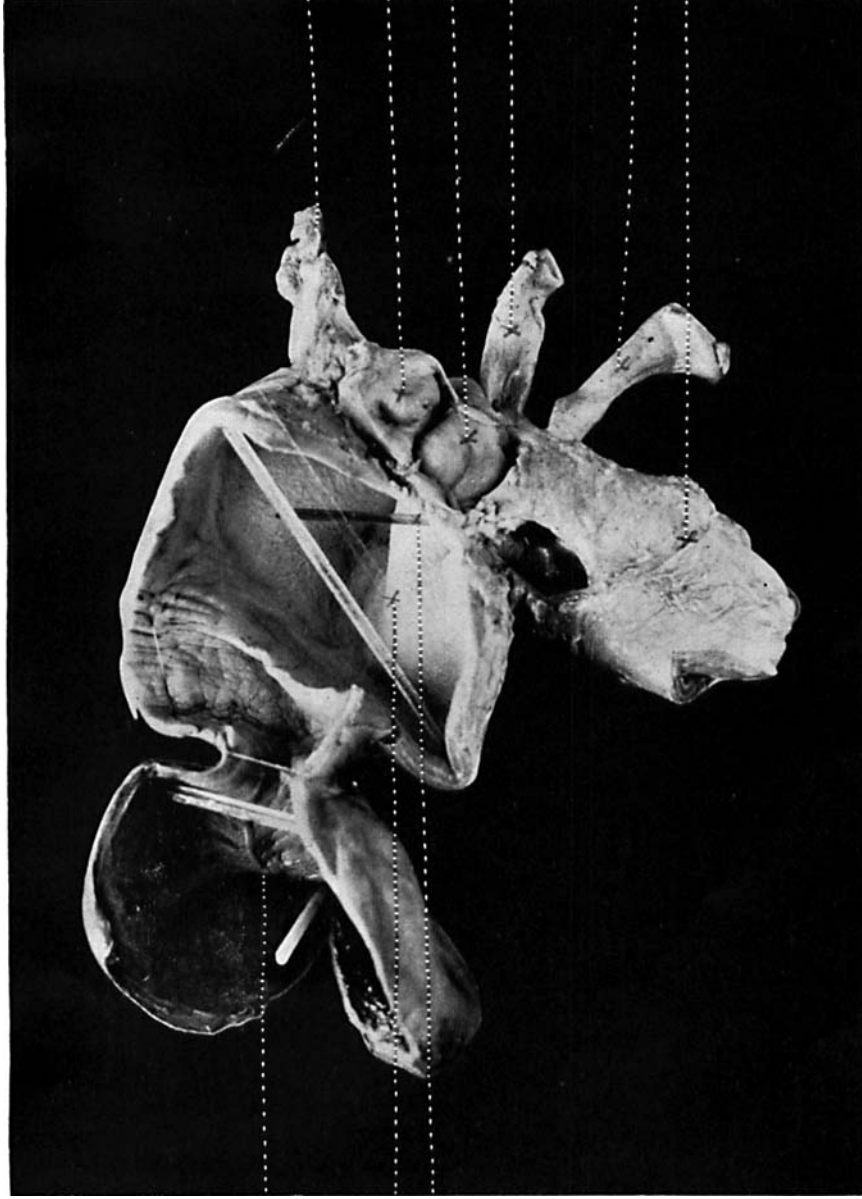


FIG. 3.—Showing bladder, right ureter, left ureter, left vagina, left uterus, left Fallopian tube, right vagina, right uterine cavity with posterior median raphe.



FIG 4.—Outer surface of perineal vesicle, covered by degenerating stratified epithelium, containing dilated glandular spaces and blood-vessels.  
No. 2 ocular,  $\frac{3}{4}$  in. objective, camera extension 40 centimetres.



FIG. 5.—Showing the inner lining of the perineal vesicle, consisting of one or more layers of epithelial cells closely adherent to the degenerated fibro-areolar stroma.  
No. 2 ocular,  $\frac{3}{4}$  in. objective, camera extension 40 centimetres.

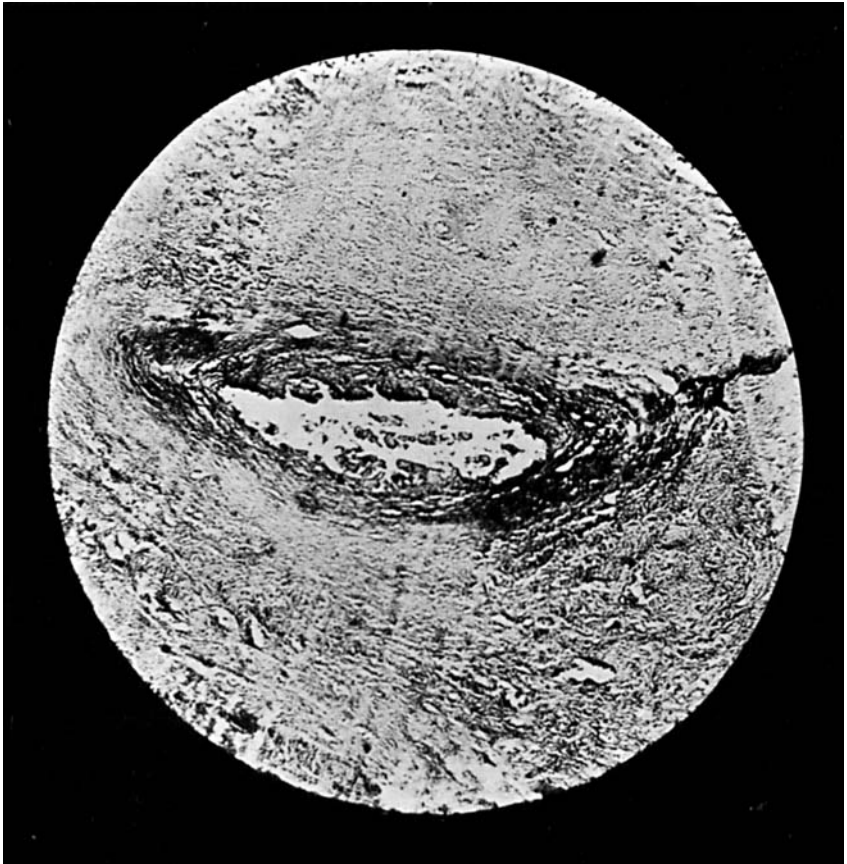


FIG. 6.—Section across the peduncle of the perineal vesicle, showing the small lumen lined by epithelium, which is supported by rather denser fibro-areolar tissue.  
No. 2 ocular,  $\frac{3}{4}$  in. objective, camera extension 40 centimetres.

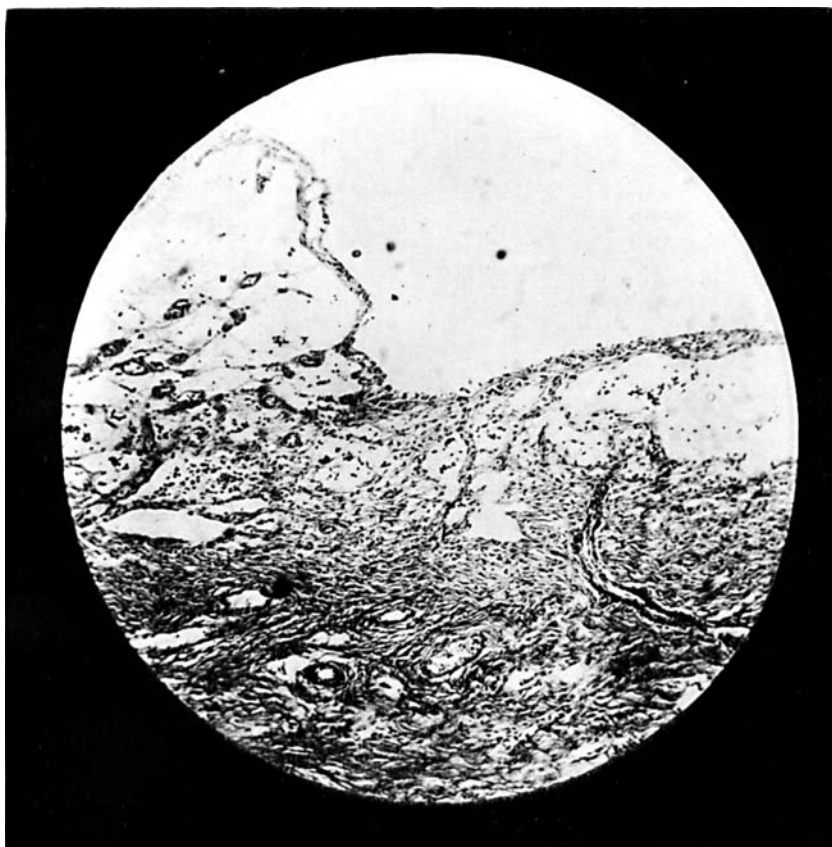


FIG. 7.—Showing the lining membrane of the right uterus (upper loculus); few epithelial cells on surface; loose areolar tissue below, rich in lymph spaces and blood-vessels.

No. 2 ocular,  $\frac{3}{4}$  in. objective, camera extension 40 centimetres.

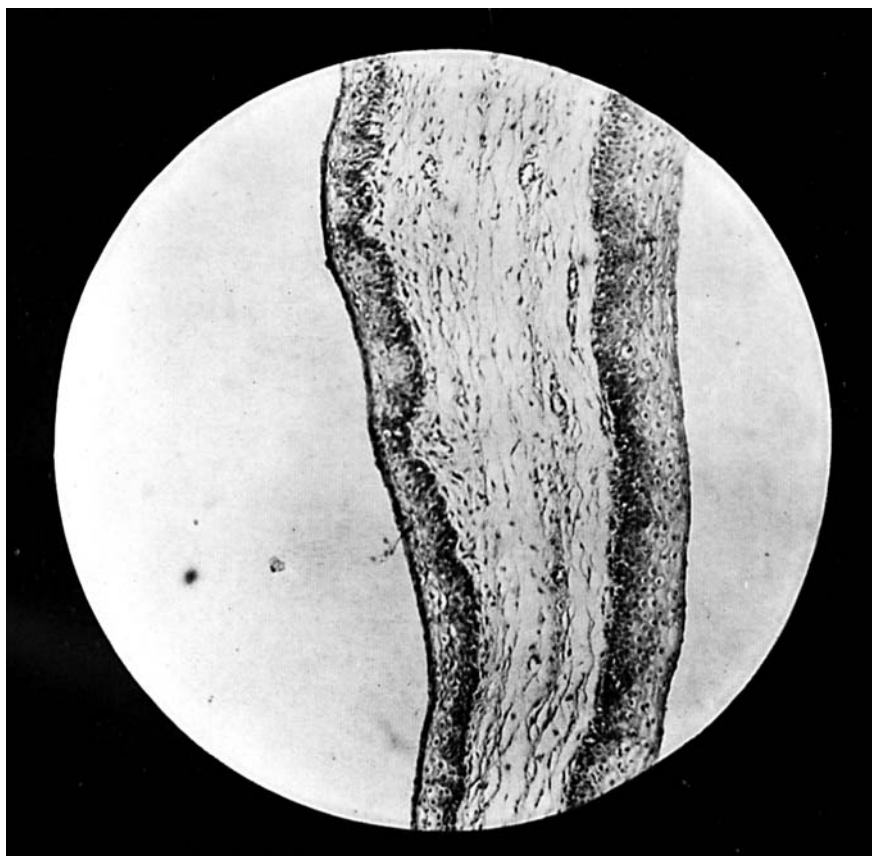


FIG. 8.—Section across the inter-vaginal septum. Left vagina to the right, and right vagina to the left, delicate fibro-areolar tissue exists between the two epithelial layers.

No. 2 ocular,  $\frac{3}{4}$  in. objective, camera extension 40 centimetres.

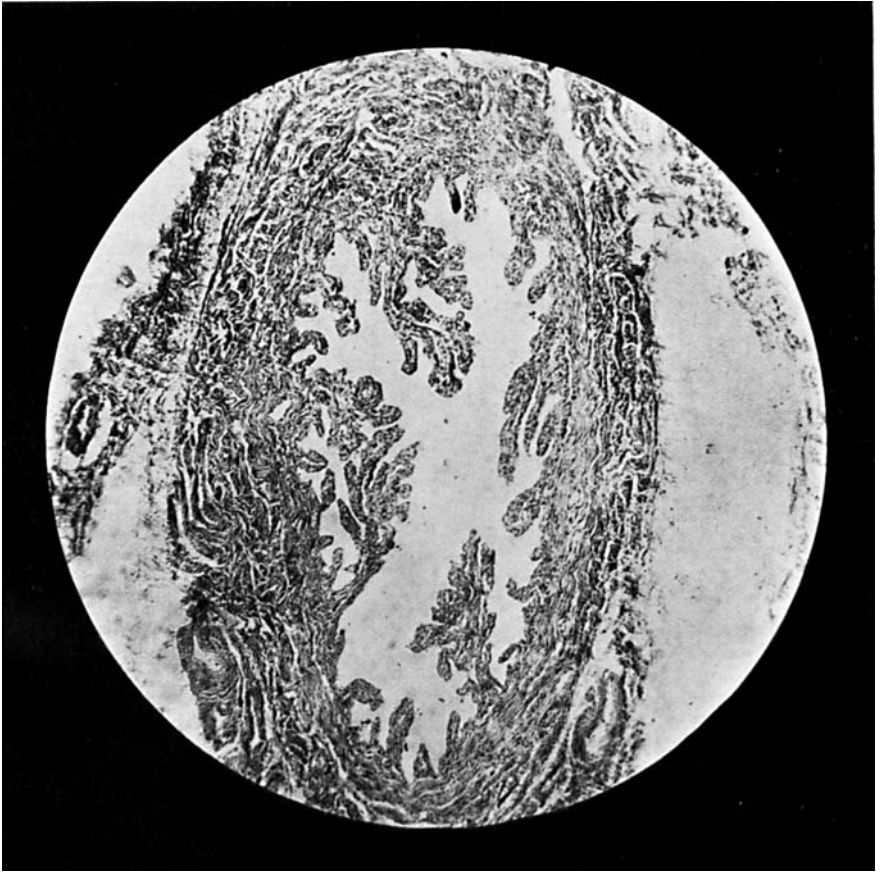


FIG. 9.—Section across the right Fallopian tube at a point one inch from its opening into the right uterus (upper loculus).

No. 2 ocular,  $\frac{3}{4}$  in. objective, camera extension 40 centimetres.

showed well-marked anterior and posterior median raphe. The left uterus and Fallopian tube were normal, the cervix possessing the usual anterior and posterior median raphe. The left vagina had well-marked fornices, containing a white creamy liquid and ended as a blind tube. Each vagina was lined by stratified squamous epithelium, and their most dependent parts came just within the pelvic brim. The rectum ended as a conical-shaped blind tube just behind the right vaginal wall, and was adherent to it. The remainder of the true pelvis was filled up by fat and areolar tissue.

*Remarks.* With our present knowledge of human embryology it is not possible to give satisfactory explanations of all the many mal-developments of the case I have recorded in detail; nor, indeed, is it my intention to attempt to indicate the probable embryological processes leading to those malformations, except in those associated with the pelvic floor and pelvic contents. Let us refer to the perineal condition first.

The normal proctodæal or primitive perineal depression is divided into an anterior or urogenital and a posterior or anal depression early in the first month of life, by the ingrowth of lateral septa. These septa uniting with the coronal septa which divide the cloaca into urogenital sinus and rectum, form the perineal body. In this foetus the proctodæal depression has not been subdivided into urogenital and anal depressions, and for the same reason there is an absence of the perineal body in the proper sense.

The cloacal membrane or plate which ought to disappear between the fourth and the sixth week, first opposite the urogenital sinus and later opposite the rectum, has in this case failed to do so, so that there are no external openings to urethra, vagina or rectum.

About the end of the second month the genital eminence and lateral genital folds make their appearance in relation to the urogenital aperture; the former becomes the glans clitoridis and the latter form the frenum and prepuce of the clitoris, the labia minora and the fourchette. All these structures are either absent or mal-developed in the case before us. This leads me to speak of the probable nature of the perineal vesicle. Two possible modes of formation suggest themselves. Firstly the lateral genital folds may have united by their inner free margins, as indeed they do in the male, and would thus cover in the urethra; a perforation of the genital eminence or glans clitoris fails to take place, thus leaving a blind *cul-de-sac* covered by labia minora and prepuce; a gradual accumulation of urine occurring in this *cul-de-sac* would be sufficient to dilate it and would tend to make it assume its pyriform shape. This explanation, of course, implies a disappearance of the cloacal membrane as far as the urogenital depression is concerned. The second explanation involves a continuance of the cloacal membrane which



indeed becomes the wall of the vesicle. Urine might accumulate in the lowest part of the urogenital sinus or urethra, and this bulging outwards the cloacal membrane gradually and slowly causes it to assume the shape of a pedunculated collection of urine. The histology of the vesicle, I think, is better understood by accepting the first explanation.

\* Dr. Berry Hart records a case of a pediculated perineal tumour in a female child, possessing the histological structure of rectum, and which was regarded by him as part of the post-anal gut. The pedicle of this tumour was attached between the vagina and the anus.

The labia majora develop as external genital folds after the third month, and these are represented in our specimen as united behind in a posterior commissure very much like the appearance seen in a male child's scrotum.

In dealing with the pelvic viscera of this case it will be an advantage to trace the probable changes in the cloacal development first.. The hind gut or cloaca begins to show signs of division into an anterior or urogenital portion and a posterior or rectal portion about the third week, the division being complete about the eighth or ninth week. This division is effected by two lateral coronal septa which unite below with the perineal septa to form the perineal body. To render this division of the cloaca a complete one there is a coronal mesoblastic dipping from above which forms in the female the recto-vaginal pouch, and in the male the recto-vesical pouch. What appears to have taken place in the case we are studying, is that the lateral cloacal septa, instead of dividing the hind gut into anterior and posterior portions, have by some malposition cut off the most dependent part in a more or less horizontal plane, and this developing into the urogenital sinus, has left the rectum high up in the pelvis. It is, of course, possible also that the posterior division of the cloaca has shrivelled, and this together with the failure of a coronal mesoblastic dipping to form the recto-vaginal pouch, is sufficient to explain the wide space between the rectum and the perineum.

In considering the developmental destiny of the urogenital sinus it is necessary to remember that in the human subject certain facts have been observed in embryos of certain ages, and that the changes of intermediate periods are still uncertain. It is fairly generally accepted, that from the urogenital sinus are developed the whole of the female urethra, all the bladder except the tip of the fundus, and possibly a part of the lower third of the vagina (Hart). At about the sixth or seventh week the Wolffian ducts open into the back of the urogenital sinus. About the same time the two Müllerian ducts lie in relation to the posterior surface of the urogenital sinus, and are

between the Wolffian ducts. According to certain authorities the Müllerian ducts open into the urogenital sinus also, and into that part which becomes the neck of the bladder. By the formation of a vesico-vaginal pouch and a mesoblastic dipping between urogenital sinus and the Müllerian ducts, these openings are carried lower and lower until they reach the perineal depression upon which they open separately.

The commencing fusion of the Müllerian ducts does not take place until the third month and is not complete until  $4\frac{1}{2}$  months. The fusion begins first at the cervix, next in the vagina, and finally in the uterine body. It is thus quite safe to say that up to  $3\frac{1}{2}$  months the Müllerian ducts open separately into the urogenital sinus. In the case before us it is quite easy to see that the Müllerian ducts have developed separately and that no fusion of the two has occurred at any point. Furthermore the opening of the left Müllerian duct into the urogenital sinus, if it ever existed, has closed again, whereas the opening of the right Müllerian duct into the urogenital sinus has been maintained and forms a congenital vesico-vaginal fistula situated just internal to the right ureteral opening, the exact relation it bears to the Wolffian duct at the sixth or seventh week. Seeing that no coronal dipping of mesoblast has occurred between bladder and vagina, and no vesico-uterine pouch formed, the two vaginæ have not been carried down to the vulva and so do not open in the urogenital depression.

It is a matter of some interest to find that both vaginæ are lined by stratified squamous epithelium even though placed so high in the pelvis. This would appear to me to still be consistent with Hart's view that the vagina is relined with epithelium derived from bud-like invaginations from the epiblastic Wolffian ducts. Whether the white, creamy fluid, in the left vagina was desquamated and degenerated epithelium, or whether it was of inflammatory origin or not, I cannot say. Another point of some interest in connection with the left Müllerian duct is, that the os-uteri of the left uterus, and the fornices of the left vagina were well formed.

We have hitherto assumed that the median raphe of the cervical canal represented the line of fusion of the two Müllerian ducts. It is therefore a matter of some surprise to find that these median raphe are present in both of these uteri in spite of the fact that each is developed from a single Müllerian duct where lines of fusion could not account for them.

Before leaving this interesting specimen I would like to say that the vesico-vaginal fistula might conceivably be of a traumatic nature, consequent upon the gradual accumulation of urine in a bladder that had no proper outlet, and which had burst into an adjoining vagina. This possibility, however, is to my mind most unlikely in face of the

tremendous hypertrophy of the right vaginal wall which, I think, indicates a very gradual distension with which hypertrophy kept pace. That it could not be an inflammatory accumulation in the right vagina which had burst into the bladder is proved by the absence of inflammatory deposit in the clear urine found, and the absence of any inflammatory appearances in the microscopical sections examined. I am, therefore, of opinion that this is a true congenital fistula which can be explained by the persistence of that primitive communication between the Müllerian ducts and the urogenital sinus which is established about the sixth or seventh week of intra-uterine life.

In conclusion I wish to thank Professor Leith, who kindly placed the material of the museum and the technique of the Pathological department at my disposal.