On malformations of the hind end of the body.

Contributors

Royal College of Surgeons of England

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ON

MALFORMATIONS OF THE HIND END OF THE BODY.

I.—SPECIMENS ILLUSTRATING MALFORMATIONS OF THE RECTUM AND ANUS.

When, in October, 1773, Hunter gave his first course of lectures on the "Principles and Practice of Surgery" at his house in Jermyn Street, introducing, as was his wont, "so much of the Animal Economy as may be necessary to illustrate these Principles," he produced in support of the truth of his teaching those preparations which afterwards formed the nucleus of this museum. On the table now before you there are a number of specimens made by Hunter to illustrate the malformations of the rectum; side by side with them are others from his "Physiological Series" to show the manner in which matter is voided from the hind end in vertebrate animals. Here are other two preparations of malformation of the rectum, made and presented by Sir William Blizard, on which he doubtless gave many a discourse to a long-past generation of students at the London Hospital. The foundation of the museum was thus made on a "teaching" collection. The magnitude of the collection now in the museum, the accumulation of material that requires an expert eye for its proper interpretation, are apt to obscure the fact that in its origin and in its growth the museum is designed not only for the purpose of preserving rare and precious specimens, but also for the purposes of teaching. In order that the unrivalled wealth of material at the disposal of this College may be the better utilized for the advantage and progress of medicine, its Council has committed to Professor Shattock and myself for the present winter session the responsible duty of bringing before you sets of specimens from the museum and explaining to you the facts they show and the principles they illustrate. Teaching is the touchstone of a museum; it discovers both the merits and deficiencies of a collection; we wish to make both known to you, so that you may assist us to improve the first and mend the second.

Altogether there are 37 specimens of malformations of the rectum in the museum, but that my description might be founded on as wide a basis as possible, I have taken the liberty of examining similar preparations in the museums of the metropolitan medical schools, in which there are 77 specimens of malformations of the rectum, the total series thus amounting to 114, which are grouped in the accompanying table. The preparations in museums are selected examples, and yet, when the series now dealt with is compared with the 100 cases collected by Curling,[1] or those tabulated by Harrison Cripps[2] some fifty years later, it will be found that the museum series is not markedly different from that which is to be expected in practice. Some of you will go through life without having to deal with a single case, for they are to be found in only 1 of 5,000 births, yet such cases will occur to many of you, and your success in treatment will depend on your knowledge of the condition. My classification differs from that of others in that sex and the relationship of the rectum to the sexual organs are made the basis of grouping. In the male that form of abnormality where the rectum opens into the urethra is made the central form round which the others are grouped; in the female, the type where the rectum opens in the vulva is made the central form (See Figs. 1 and 5).

Malformations of the Rectum in Males.

Group I.—*Males in which the Rectum opens into Urethra.*

In Fig. 1 is represented the typical form of this abnormality; the orifice of the rectum is situated at the lower end of the veru montanum, being thus immediately beyond the opening of the uterus masculinus and vasa deferentia. It is valvular in form. In 4 cases the orifice was between 2 and 3 mm. in diameter; in 5 between 1 and 2 mm.; and in 6 merely large enough to take a bristle, therefore too small to allow the passage of faeces. In all specimens that have been laid open the orifice is seen to be provided with a sphincter. Of 22 cases in which it was possible to see the exact position of the orifice, it was found to occur at the lower end of the veru montanum in 17; in 1 it was at the apex of the prostate, in 1 by the side of the veru montanum, in 1 at the internal meatus, and in 1 in the trigone of the bladder. In all of the College museum specimens the orifice is in the typical position. Curling estimated that this form made up rather more than a fourth of cases seen in practice; 60 per cent. of museum specimens belong to this type.

Group II.—*Males in which the Rectum ends as a Fibrous Cord at or above the Base of the Prostate.*

Of this group there is not a single specimen in the College collection; but there are seven examples in other metropolitan museums. From Fig. 2 it will be seen that

Group II represents a greater degree of arrest of development, or of atrophy, of the rectum than is shown in Group I.

TABLE I.—*Specimens of Malformation of the Rectum in London Museums.*

Groups.	In Museum of Royal College of Surgeons.	In Museums of Metropolitan Medical Schools.	Total
A. MALES (see Fig. 2):			
1. Rectum opening in urethra ...	7	26	33
2. Rectum ending as cord at or above base of prostate	0	7	7
3. Rectum ending as cord at site of proctodaeum	2	5	7
4. Rectum ending blindly at proctodaeum	1	6	7
B. FEMALES (see Fig. 4):			
1. Rectum ending in vulva or vagina	1	5	6
2. Rectum ending in cord above upper fornix of vagina	0	5	5
3. Rectum ending as cord at upper fornix of vagina	0	3	3
4. Rectum ending as cord on vagina below upper fornix	0	2	2
5. Rectum ending blindly or as cord at site of proctodaeum	3	7	10
C. MISCELLANEOUS SPECIMENS:			
1. Imperfect	1	5	6
2. Imperforate rectum in females with male form of external genital organs	3	3	6
3. Rectum opening abnormally ...	1		1
4. Rare malformations	2	3	5
5. Abnormalities of the rectum in domesticated animals	16	0	16
	37	77	114

GROUP III.—*Males in which the Rectum terminates in a Fibrous Cord at the Site of the Proctodaeum* (Fig. 2).

Of this group there are two examples in the College collection, and five in the school museums. The nature of this malformation and its relation to the other groups will be apparent when the embryology and evolution of the rectum is dealt with.

GROUP IV.—*Males where the Rectum ends Blindly at the Proctodaeum* (Fig. 2).

There is one specimen of this form in the College collection, and six in the school museums, several of which are of great interest, especially two in the museum of St. Bartholomew's Hospital Medical School (preparations 3647 and 3648), and a third in the museum of Guy's Hospital Medical School (preparation 983). The two preparations first named were obtained from children aged 14 months and 24 months, who had been operated on at birth. A ring marks the site of the congenital septum, and in one of them the mucous membrane of the rectum has spread downwards some distance into the anal canal after perforation.

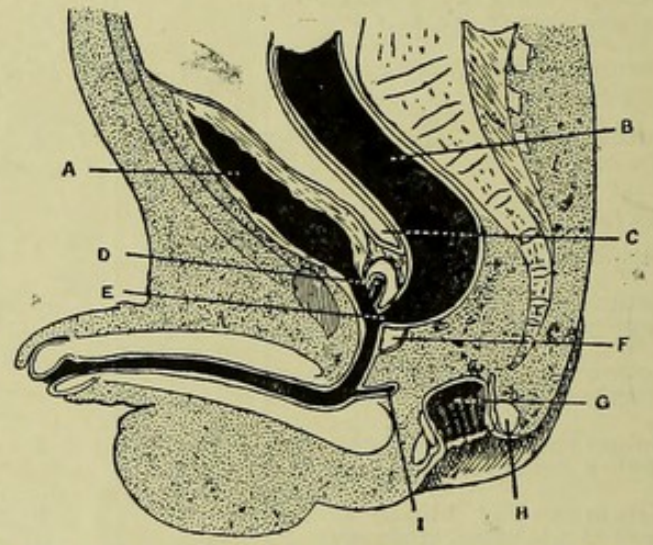

Fig. 1.—Section of pelvis of a male chjld, showing the rectum opening into the prostatic part of the urethra. A, Bladder; B, rectum; C, recto-vesical pouch; D, uterus masculinus; E, intracloacal anus; F, prostate; G, proctodaeum; H, external and internal sphincters; I, Cowper's gland.

THE NATURE OF RECTAL MALFORMATIONS.

How is such a series of malformations to be explained? The usual explanation given is that they are the result of an imperfect division of the cloaca. In the series of malformations now described to you there is not one single form that is satisfactorily accounted for thereby. The modern tendency is to explain all abnormalities on a purely embryological basis; we are only beginning to realize the fact that the earlier processes of development of the mammalian embryo are profoundly modified to serve the purposes of the intrauterine life, and that it is only by appealing to the facts of the "animal economy," as Hunter did, that we may hope to interpret rightly the processes observed by the embryologist. This is especially true of the hind end of the body, parts of which, such as the allantois, have been transformed for the early nourishment of the embryo. The only serious attempt which has been made to explain the abnormalities of the rectum is

that of Dr. F. Wood Jones.[3] The urethral orifice of the rectum he regards as the original anus; the part of the rectum between that opening and the perineal anus—which he regards as a new orifice—is a post-allantoic prolongation of the original rectum. To myself, Wood Jones's theory was at first quite satisfying; it explained very well the numerous abnormal forms met with; it was only when tested afterwards by a study of the evolution of the rectum in various vertebrate forms, and with a more complete knowledge of the development of the rectum, that it seemed to me to break down. The key to these abnormalities, I think, is to be found in a study of the comparative anatomy and physiology of the cloaca, rectum, and urino-genital passages. Above all, the process of impregnation

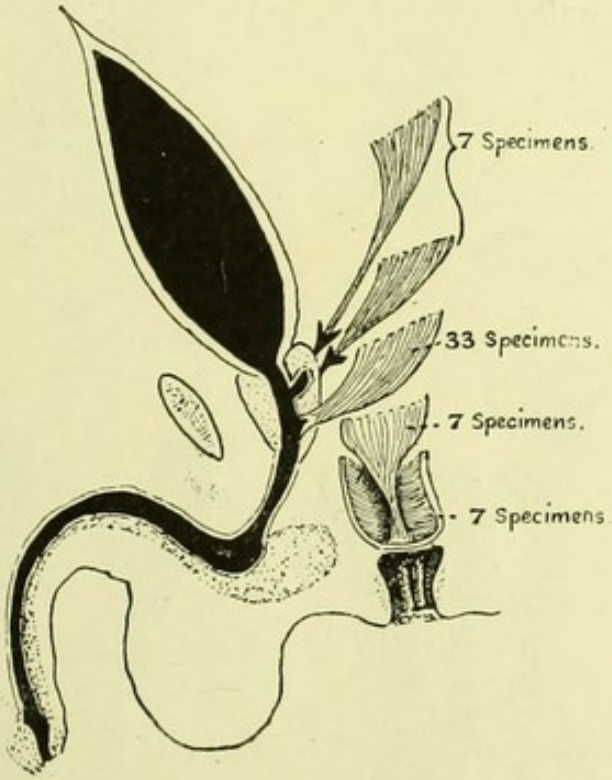

Fig. 2.—Illustrating the various degrees of imperfection of rectum in the male. The rectum is shaded; the various degrees of imperfection make up a series between the proctodaeum and base of the prostate.

has to be kept in mind, for it was by the evolution of the penis that the rectum attained an opening on the perineum. In Fig. 3 stages in the evolution of the mammalian form of perineum are exemplified, and on the table are specimens representing those stages. Much of the obscurity regarding the hind end of the body results from an imperfect knowledge of what the cloaca is. The cloaca of the frog is represented in the mammal by the trigone of the bladder together with the urethra; it is a closed passage that at times serves to convey the urine and genital products to their destinations; the faeces have no lodgement there, only a right of passage. It will be observed from Fig. 4 A that the rectum and ducts of the

kidney and genital glands open on the dorsal surface of the cloaca, but the rectum perforates higher up, further from the tail, than the genital and urinary ducts. In the 33 specimens of malformed rectum there was only one which opened in the trigone of the bladder, but that one

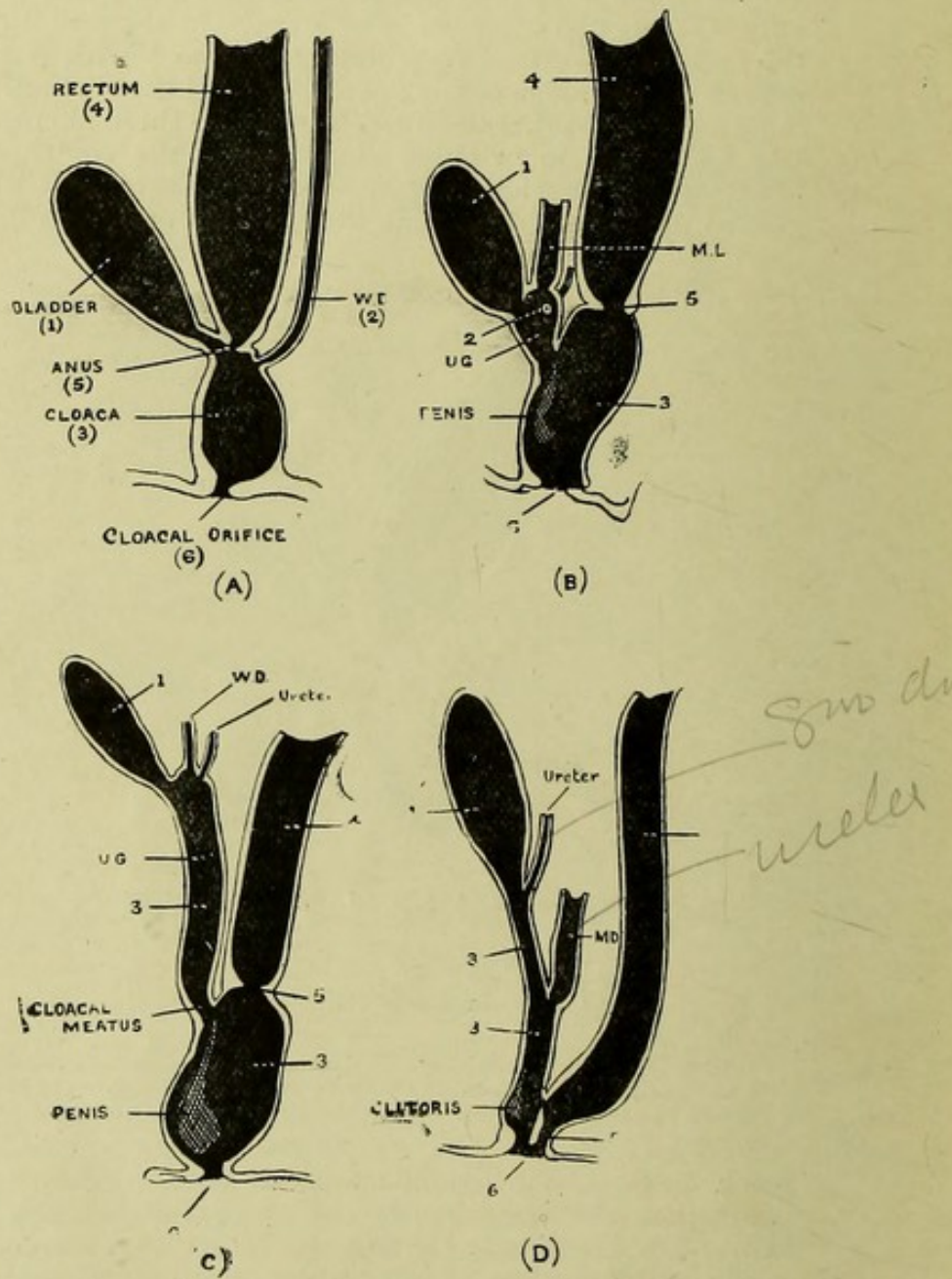

Fig. 3.—Diagrams to show the manner in which the cloaca is modified and the termination of the rectum transferred from the cloaca to the perineum in higher vertebrates. (A) the amphibian form: (1) bladder; (2) Wolffian duct (ureter and vas); (3) cloaca; (4) rectum; (5) intracloacal anus; (6) cloacal orifice. M.D, Mullerian duct; (B) form found in the tortoise; (C) form in Monotremes; (D) form found in female marsupial.

reproduced exactly the amphibian form. In the tortoise and turtle, in which are foreshadowed so many mammalian structural adaptations, a further step is found (Fig. 4 B); the rectal orifice has moved along the dorsal wall of the

cloaca so that it now opens nearer the tail than the genital and urinary ducts; it now opens exactly in the position where the majority of specimens shows the abnormal human rectum to open—in the floor of the urethra just beyond the veru montanum. In

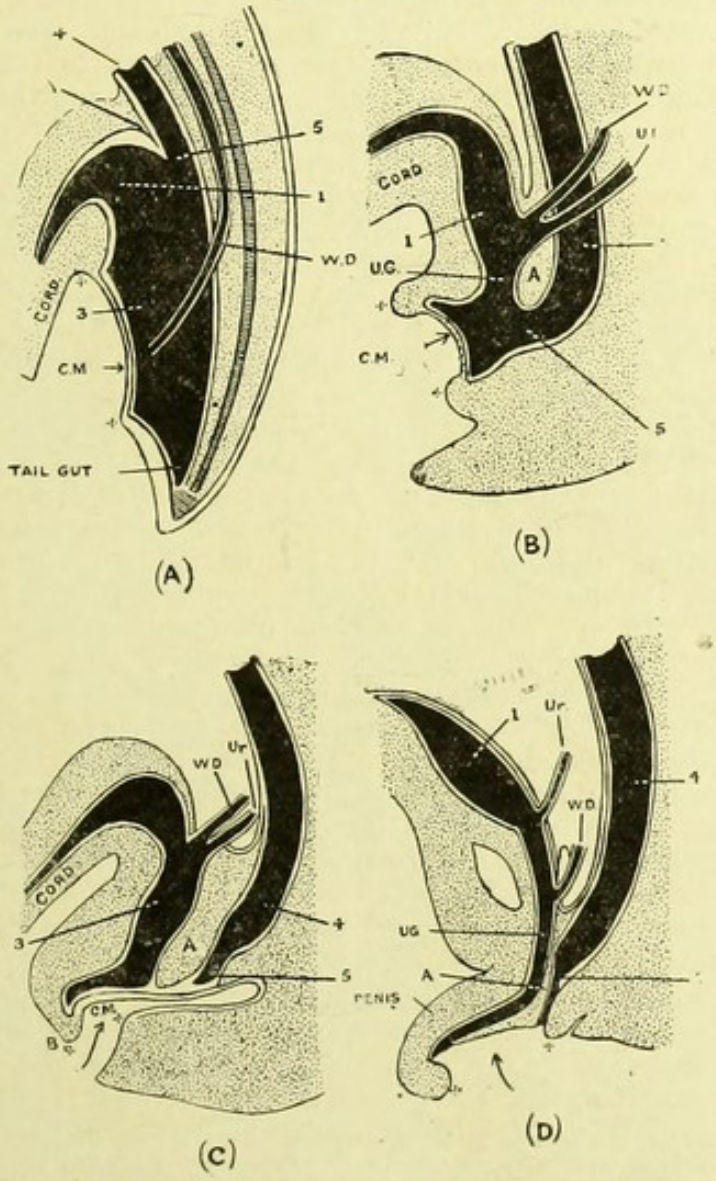

Fig. 4.—Showing the manner in which the rectum becomes separated from the urogenital sinus (endodermal cloaca) during development of the human embryo. (A) From human embryo, 4 mm. long (about twenty days); after Keibel. (B) From human embryo, 11 mm. long (about thirty-five days); after Keibel. (C) and (D) Later stages of development: 1, Bladder; 2, Wolffian duct (ureter and vas); 3, endodermal cloaca; 4, rectum; 5, anus; C.M, cloacal membrane; U. G, urogenital sinus; A, mesoblast at junction of rectum and endodermal cloaca; B, penis; XX, the limits of the perineal depression (ectodermal cloaca).

the 33 specimens one opened in the trigone of the bladder (the amphibian form), one at the meatus of the urethra, one in the sinus prostaticus; the two last named are examples of arrest in the passage from

the amphibian to the tortoise form. The group of cases where the rectum ended in a cord on the base of the prostate also represent stages of arrest in passing from the amphibian to the tortoise condition, but in them the arrest was accompanied by an atrophy of the terminal part of the rectum, thus producing a fibrous cord.

To understand the nature of those malformations where the rectum ends blindly at the proctodaeum (see Fig. 2) one must take into consideration the changes in form and function which are seen to occur in the reptilian and lower mammalian cloaca. In the tortoise and turtle the cloaca is becoming modified for sexual purposes; on its ventral wall a phallus or glans has been formed (Fig. 4 B); it is grooved for the conduct of the seminal fluid; the cloaca has become elongated and capable of eversion and intromission. With these sexual modifications the cloaca becomes less capable of serving as a faecal passage, and hence the orifice of the rectum is moved nearer to the perineum. In the Monotremes the sexual modification is carried a stage further; the rectum now opens into that part of the cloaca derived from the ingrowth of ectoderm —the ectodermal cloaca. The deeper part of the cloaca— the part derived from endoderm—now serves solely as a urogenital passage and is known as the urogenital sinus. The endodermal cloaca becomes the urogenital sinus. In marsupials, and in mammals generally, the cloaca* has become more and more adapted for a purely genital function; in the male it becomes elongated, everted, and its mouth prolonged with the outgrowth of the penis. These modifications will be more fully described in connexion with hypospadias. During the evolution of the higher vertebrates the anus has migrated from an intracloacal to an extracloacal or perineal position. The various forms of malformations of the rectum represent arrested stages of migration (see Fig. 2).

In Fig. 4 are represented stages in the development of the human rectum. It is seen to be gradually separated from the cloaca; the embryologist interprets the appearances to mean that the cloaca is being divided into a dorsal part, which becomes the rectum, and a ventral part, which forms the urogenital sinus. One cannot easily conceive the transformation of a part of the cloaca into a part of the rectum; they are so radically different in structure and function. Nowhere in the animal kingdom is there evidence of such a change. It is clear, I think, that the proper interpretation of Keibel's models is not that the cloaca is being divided, but that the rectum is gradually migrating from the amphibian to the mammalian position, and may undergo arrest at any stage of its progress during the development of the embryo.

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* The term "cloaca" is used throughout in its primitive sense—the common passage for rectum, bladder, and genital ducts. The embryologists have sought to restrict the term to that part of the cloaca derived from endoderm (hind gut).

Cause of the Abnormalities.

The key to these abnormalities is likely to be discovered from experiments made on developing animals. Professor Mall [4] has lately published an account of the examination of 163 malformed human embryos, and in the chorion of every one of them he found evidence of uterine inflammation. Uterine inflammation will be found to be the commonest cause of fetal malformation. One must keep in mind, too, that the migration of the rectum is connected with a sexual process—namely, the evolution of a penis and transformation of the cloaca to form a genital passage. It is, therefore, possible that the cause of arrest may be found in a primary defect of the sexual glands.

Malformation of the Rectum in Females.

Group I.—*Females in whom the Rectum Opens in the Navicular Fossa of the Vulval Cleft* (see Fig. 5).

This abnormality in the female corresponds exactly to the form in the male where the rectum opens in the

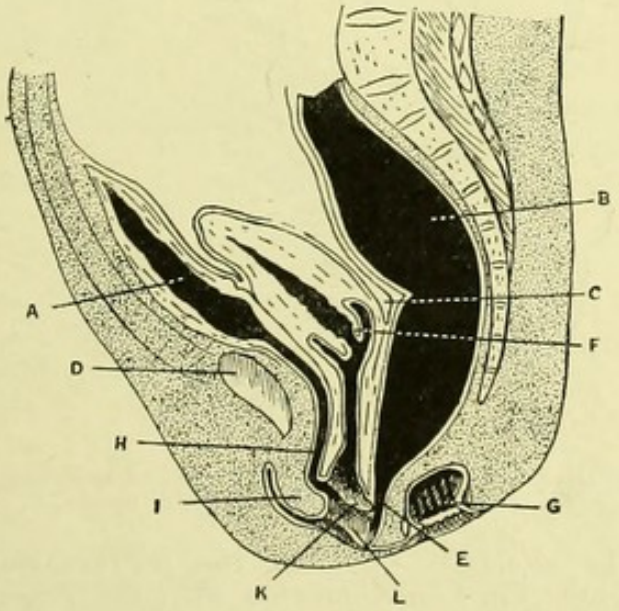

Fig. 5.—Section of pelvis of female infant, showing the rectum opening into the navicular fossa of the vulva. A, Bladder; B, rectum; C, recto-uterine fold; D, symphysis; E, vulval anus; F, cervix; G, proctodaeum (rarely present if rectum opens in vulva); H, urethra; I, clitoris; K, hymen.

floor of the urethra at the lower end of the veru montanum. In both sexes the opening is situated immediately below the opening of the genital ducts into the cloaca. In the female human embryo the cloaca undergoes a transformation in connexion with the development of the vagina and uterus; Berry Hart [5] and also Wood Jones [3] have described these changes. After the rectum has migrated along the cloaca to its perineal termination, the cloacal ends of the Müllerian ducts become filled with solid rods of cells, and burrow and press downwards, to open nearer the perineum in that part of the cloaca which

forms the vulval cleft. In the female the original cloaca is transformed into (1) the trigone of the bladder, (2) the urethra, (3) the space between the labia minora. In the College museum there is only one specimen of the rectum opening in the vulval cleft; there are five in the school museums. Of the 6 cases from which these preparations were derived, 2 died at birth, 1 died at 2½ years of age, another at 5½ years, another at 11 years, the remaining being an adult. Bardeleben[6] has recently tabulated notes on 31 such cases observed chiefly in adults. A case has been well figured and described lately by Mr. F. S. MacKenzie.[7] Of all the forms met with the prognosis is by far the most favourable in this class. In one case the opening seemed to be in the vagina, but as no hymen was present it was impossible to identify with certainty the junction of vagina and vulva.

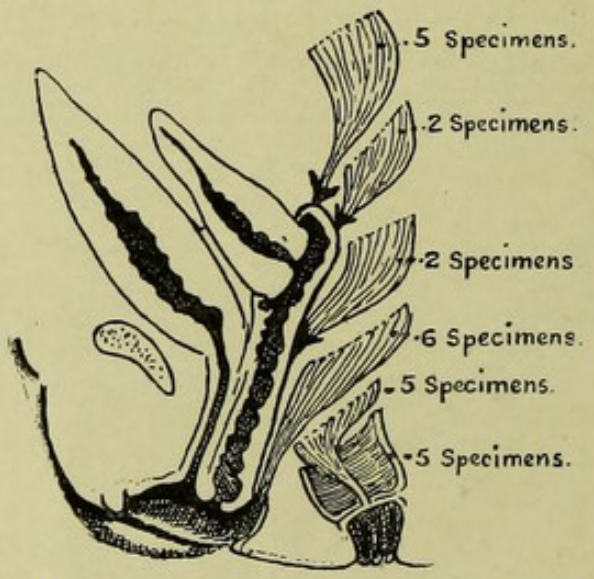

Fig. 6.—Diagram to show the termination of the rectum in 25 female infants (see explanation in text).

GROUP II.—*Females in whom the Rectum ends as a Fibrous Cord in connexion with the Vagina.*

There are ten examples of this condition in the London school museums, but not one in the College museum. They represent (see Fig. 6) arrest of development in the migration of the rectum along the cloaca to the perineum. In some recorded cases the rectum is said to have opened into the vagina. In such cases as I have examined the vagina has been divided, the arrested rectum probably preventing the proper fusion of the Müllerian ducts to form the vagina.

GROUP III.—*Females in whom the Rectum ends Blindly or as a Fibrous Cord at the Site of the Proctodaeum.*

As may be seen from the table on page 1737, there are ten examples of this malformation, three of them being in the College collection. As in the male, this form represents

stages of arrest in the passage of the rectum from its cloacal to its perineal position.

It will be observed that the cloaca undergoes less transformation in the female than in the male.

Miscellaneous Specimens.

The two series of abnormalties just described embrace most of the forms of imperfect rectum, but there are also a number of uncommon anomalies which are best grouped together (see Table on page 3).

1. *Imperfect Specimens.*—In every museum there are to be found certain preparations which are so imperfect that it is impossible to tell what the condition was in life. These are merely mentioned here to draw attention to the fact that it is necessary to examine and retain the relationships of the rectum to all the parts of the pelvis if a perfect record is to be secured. In the six specimens which are classed here as imperfect, only the conical termination of the rectum being preserved, one has to guess the relationship that obtained during life.

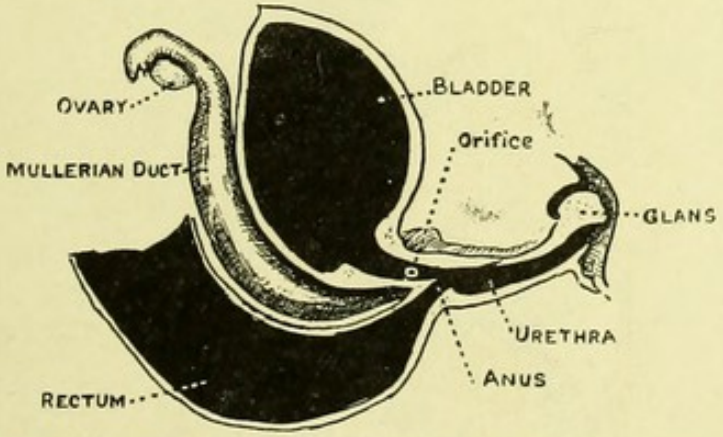

Fig. 7.—Female with rectum opening into urethra completed by clitoris (from specimen in College collection).

2. *Females with Clitoris Simulating a Penis and with the Rectum opening into the Urethra* (Fig. 7).—This group of cases is best dealt with under malformations of the external genital organs. There are three specimens in the College collection and three others in the school museums; all six are exactly of the same form; in all the vulva and clitoris are prolonged to form a urethra, the rectum opening in the floor of the urethra immediately below the orifice of the vagina. The abnormal rectal opening in this group is exactly similar to that seen below the veru montanum in the male, and in the vulva of the female.

3. *Abnormal Position of the Anal Orifice.*—In describing the development and evolution of the mammalian rectum, reference was made to: (1) The migration of the rectal orifice from the cloaca to the perineum; (2) to the prolongation forwards of the perineal orifice of the urogenital sinus in the male to form the urethra. Now it occasionally happens that not only is the perineal orifice of the cloaca carried forwards on the out-growing penis, but the rectal

orifice is also transplanted forwards with it. Fig. 8 will serve to make the matter clear. Esmarch, and later Stieda,[8] have described cases where the rectum opened in the frenum of the prepuce; it may open on the under surface of the penis, or on the raphe of the scrotum, as is shown by a specimen in the College museum, the only one of this class I have been able to examine; or it may open on the perineum. The perineal raphe, it will be remembered, stretches from the normal anus to the frenum of the prepuce; the termination of the rectum may, in these abnormal cases, be found at any point in the median raphe. These abnormalities can only occur in males; the developmental process which causes the prolongation of the cloaca to form the urethra in these cases extends also to the rectal orifice. The explanation of such

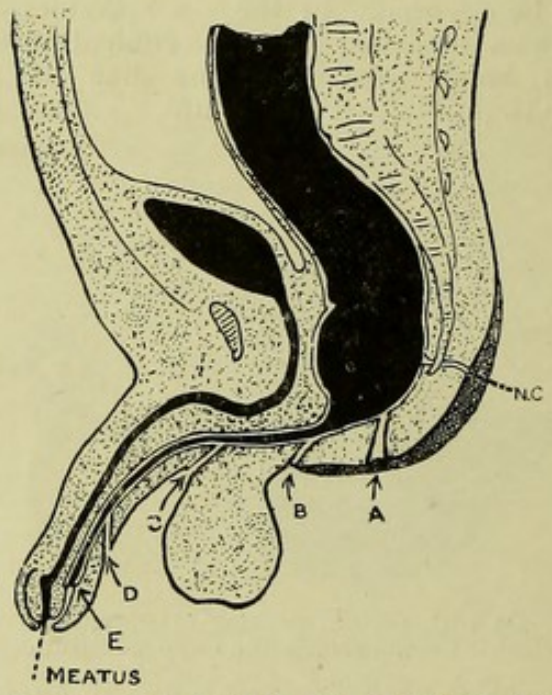

Fig. 8.—Showing the normal and abnormal sites of anus. A, Normal position; B, perineal orifice; C, scrotal orifice; D, subpessile; E, intrapreputial; N.C., neurenteric canal.

will be best given in connexion with hypospadias. Occasion will be taken then to mention those peculiar cases, of which there is no example in the College collection, where the median scrotal raphe is continued back as a free band across the anal orifice to join with the perineal tissues between the anus and the coccyx, as recently described by Bate[9] and by Kelly.[10] The specimen in the College collection (No. 765) is unique in that the rectum has two openings: (1) a proctodaeum has grown in and opened into the rectum, thus forming an anus in the usual situation; (2) the cloacal orifice has been prolonged forwards, and opens in the median raphe of the scrotum near the root of the penis. In Fig. 8 the position of another rectal opening or fistula is shown near the point of the coccyx. Curling observed the escape of

faecal matter by such an opening—the neurenteric part of the blastopore. There is no example of this condition in the museum.

4. *Rare Malformations.*—Under this heading are classified a number of very uncommon conditions. Preparation No. 16 of the Teratological Series shows an arrest of the cloaca at an early embryonic stage; in No. 649 B the rectum is absent and the proctodaeum has opened into the urethra; in a preparation in the London Hospital Medical College Museum practically the whole of the colon is absent; in the school museum of Guy's Hospital a preparation (No. 983) shows that the proctodaeum has opened into the ventral surface of the rectum in place of into its caudal end; in this case ectopia vesicae with compression of the pelvis were present. Evidently there had been an early disturbance of parts, so that the normal relationship between proctodaeum and rectum had been altered. In No. 558 of the College collection, a specimen presented by Mr. Curling shows a septum across the rectum about 4 in. from the anus. It is the only specimen of its kind known to me, and it is not easily explained. In the Museum of St. Bartholomew's Hospital is a rectum (No. 3648 B) with a cyst attached to its posterior wall; it is exactly in the position and of the form one would expect a derivative of the post-anal gut to occupy.

5. *Abnormalities of the Rectum in Domesticated Animals.*—Of the 16 specimens in the College museum showing abnormalities of the rectum in animals, 8 belong to one type which shows a forking of the rectum with two anal orifices, usually associated with a corresponding dichotomy of all the parts of the hind end of the body, penis, bladder, etc. Posterior dichotomy is represented by 4 specimens derived from birds and 4 from mammals (1 pig, 3 dogs), but we have no human example of this abnormality in the museum. The 8 remaining specimens—of which 6 are derived from the pig, 1 from the sheep, and 1 from the dog—show abnormalities of the rectum and anus similar to those described in man. In 3 sows the rectum opens into the vulval cleft; in another, a female with external genitals of the male type, the rectum opens in the urethra below the vagina. In two male animals the rectum terminates, or has terminated, in the bulbous part of the urethra; there are two specimens in which the proctodaeum is imperfectly formed and the rectum unopened. In one specimen the uterus appears to terminate in the last part of the rectum.

Development of the Proctodaeum.

Many preparations of imperforate rectum show very definitely that the anal canal with the internal sphincter and external sphincters arises in connexion with the perineum and independently of the bowel. One scarcely expected that such could be the origin of the internal sphincter, but there can be no doubt about the matter; the evidence of the specimens is so definite. The rectum

proper—the part derived from the hind gut—ends at the entrance to the anal canal just above the valves or folds which Sir Charles Ball has described as joining the upper ends of the columns of Morgagni; the lower ends of the columns are also united by folds or valves; the white line of Hilton occurs just below the columns of Morgagni and marks the junction of the skin with the mucous membrane of the anal canal. Thus the columns of Morgagni, the valves joining the columns above and below, the mucous membrane of the anal canal, the external and internal sphincter are derived from the proctodaeum or perineal ingrowth. Unfortunately, in many cases no care has been taken to preserve the perineum, but the material at my disposal warrants the statement that the condition of development of the anal depression is an uncertain index of the degree to which the rectum is developed. In 7 females in which the rectum opened in the vulva the proctodaeum was absent in all; in 10 in which the rectum ended at or near the perineum, an anal depression was present in all; in 10 cases where the rectum ended blindly at or above the vagina, the proctodaeum was well developed in 7, partly developed in 1, and absent in 2. Amongst males with the rectum opening into the urethra there were 7 with the proctodaeum well marked, 3 with it partly developed, and an uncertain number in which it was absent. In 7 males in which the rectum terminated blindly at or above the prostate the proctodaeum was present in 4, absent in 3; where the rectum nearly reached the perineum the proctodaeum was well marked in all cases, partly developed in 2, and absent in 1.

In 2 cases, both females, the epithelium of the proctodaeum had budded out irregularly, so as to form diverticula, some of which projected for some distance into the perineal tissue, and undoubtedly correspond to certain diverticula which are found in the anal canal opening under the folds which join together the upper ends of the columns of Morgagni, and which were first pointed out to me by Mr. J. Bernard Dawson.

Associated Malformations.—The following is a list of the lesions associated with imperfect rectum in the human specimens examined for the purposes of this demonstration:

Retained testis	1
Malformed coccyx	2
Imperforate urethra	2
Dilated bladder	5
Low implantation of ureter	3
Occlusion of ureter	1
Urachal fistula	1
Hypospadias	1
Ectopia vesicae	1
Talipes	1
Cord to coccyx	1
Right arm deformed	1
Compressed pelvis	2

Results of Operation.

I do not propose to deal with the operative procedures to be adopted for the treatment of malformation of the rectum, but a study of these specimens has forced several matters on my attention. One is the great danger of using a trocar for the relief of the condition; there are eleven preparations mounted purposely to show perforations into the pouch of Douglas or a separation of the coats of the rectum by the use of the trocar; Curling and Harrison Cripps have emphasized the danger of the trocar. There should be no half measures in operating; in 95 per cent. of cases it is physically possible to bring the rectum down to the perineum; if it be not found terminating at the site of the proctodaeum, it will be found perforating the prostate or the lower end of the vagina; if not there, then at the base of the prostate or upper part of the vagina. If unrelieved, death occurs about the fifth day from gangrene and rupture, usually towards the upper part of the colon. The prognosis is bad; an imperforate condition of the rectum appears to react on the economy of the child even before birth, and death commonly ensues, even if the rectum be opened early. The prognosis is favourable in the case of females where the rectum opens in the vulva; in all cases hypertrophy and dilatation of the rectum follows, even if an anus is successfully established. It is important to remember that the external sphincter and also internal sphincter are developed in connexion with the perineum, not with the bowel; hence, if a proctodaeum be present, the surgeon may rely on obtaining an orifice provided with competent sphincters.

[Besides the authorities cited below, the following works may be consulted with advantage: Ballantyne's *Antenatal Pathology;* Dr. William Wright's Report on Recent Teratological Literature, published annually in the *Journal of Anatomy and Physiology; The Rectum and Anus*, by Sir Charles Ball.]

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II.—HYPOSPADIAS AND ALLIED CONDITIONS.

The specimens for to-day's demonstration illustrate a subject in which Hunter's knowledge was a hundred years and more ahead of his time. On the table are his celebrated preparations of the sexual organs of "free martins," of the foal-ass, of the sheep from the West Indies, and others described in his writings. The anomalous condition seen in these and similar preparations he ascribed to an absence of the "stimulus of perfection," which the fully normally formed sexual organs exercise on the development and physical health of the body. The phrase "stimulus of perfection" has been criticized as vague and unmeaning by many of Hunter's commentators, but to us of a later generation, who recognize the influence which internal secretions of various organs exert on the physiology of the body, it has a very definite significance. He even asserted that "the penis, the urethra, and all the parts connected with them are so subservient to the testicles that I am persuaded few of them would have existed if there had been no testicle" (vol. iv, p. 29, Palmer's edition). We are quickly coming round to Hunter's way of thinking. In recent years Professor Shattock[11] has shown that when the aged hen puts on the feathers and manners of the cock—a transformation which you can see in specimens preserved by Hunter—her ovaries have assumed certain testicular characters, and we may suppose are then capable of throwing into the circulation an internal secretion similar to that of the testes. In the third month of fetal life the sexual features are being differentiated. Professor Arthur Thomson[12] has shown that the pelvis then takes on its special sexual characters; we infer that the sexual glands must be then acting on the whole organism. It was clearly Hunter's opinion that the primary cause of all malformations of the sexual parts of the body is a temporary or permanent imperfection of the sexual glands. It is a matter of regret that the condition of the sexual organs in many of the preparations now shown has not been determined microscopically, but that is a fault which may be mended soon.

Instead of now showing and describing the various specimens illustrating hypospadias and allied conditions,

I propose first to bring before you preparations from the comparative anatomy series—many of them prepared by Hunter—which illustrate steps in the evolution of the penis and vulva, so that, when we come to discuss the malformations of these parts, the significance of their various features may be perceived. The evolution of the external genital organs of man and woman necessitated a complete transformation of the cloaca. The Edentates (sloths, ant eaters) and some of the Rodentia show an interesting stage of the process. In the ant-eater the cloacal lips are widely open (see Fig. 9); the orifice of the urogenital passage (endodermal cloaca) is seen to protrude between the lips of the cloaca, leaving the anal orifice in the depth of the cloaca. The urogenital passage is being separated from the faecal outlet and carried beyond the cloacal orifice.

In the tortoise or turtle, in Echidna (the Australian porcupine, one of the lowest mammalian forms), and in a marsupial, such as the kangaroo, are seen certain stages in the evolution of the penis. In the tortoise (Fig. 10 A) the

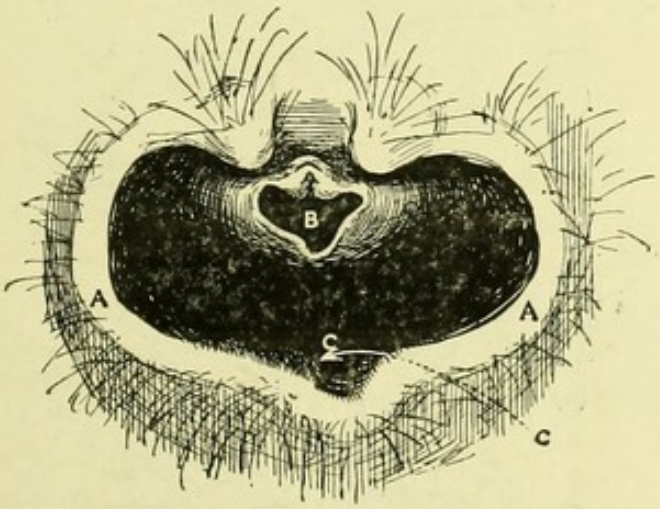

Fig. 9.—Perineum of female ant-eater (*Manis longicauda*), showing the urogenital passage being protruded from the cloacal orifice, leaving the orifice of the rectum (C) in the depth. A. A, Margins of cloaca; B, urogenital orifice; C, over anus (Dr. Stanley Beale).

penis is intracloacal; it is formed by a modification of the ventral or pubic wall of the cloaca. Practically only the glans is free; this is cleft and forms a groove; when erect the margins of the groove approximate and form a canal. When the glans is extruded during copulation, the cloacal walls contract and complete the urogenital passage; the musculature of the cloaca serves to ejaculate the contents of the canal. Thus, there is a condition of hypospadias, but the contracted walls of the cloaca supplies the blank in the floor of the temporary seminal passage. In Echidna (Fig. 10 B) a further stage is found. The penis is still intracloacal, but the lips of the glans have fused and formed what may be named the phallic canal. The seminal canal is thus made up of two parts: (1) the urogenital sinus derived from the endodermal

cloaca; (2) of the phallic canal made by union of the lips of the phallic groove. In copulation the glans and part of the penis are extruded, and the hiatus in the floor of the seminal canal (the primitive urogenital orifice) is temporarily closed. In the marsupial the penis is still intracloacal, but is capable of being almost completely extruded. The two parts of the seminal canal are united to form a permanent urethra; by their union the primitive orifice of the urogenital sinus, occluded only during copulation in Echidna, is now permanently shut. The phallic

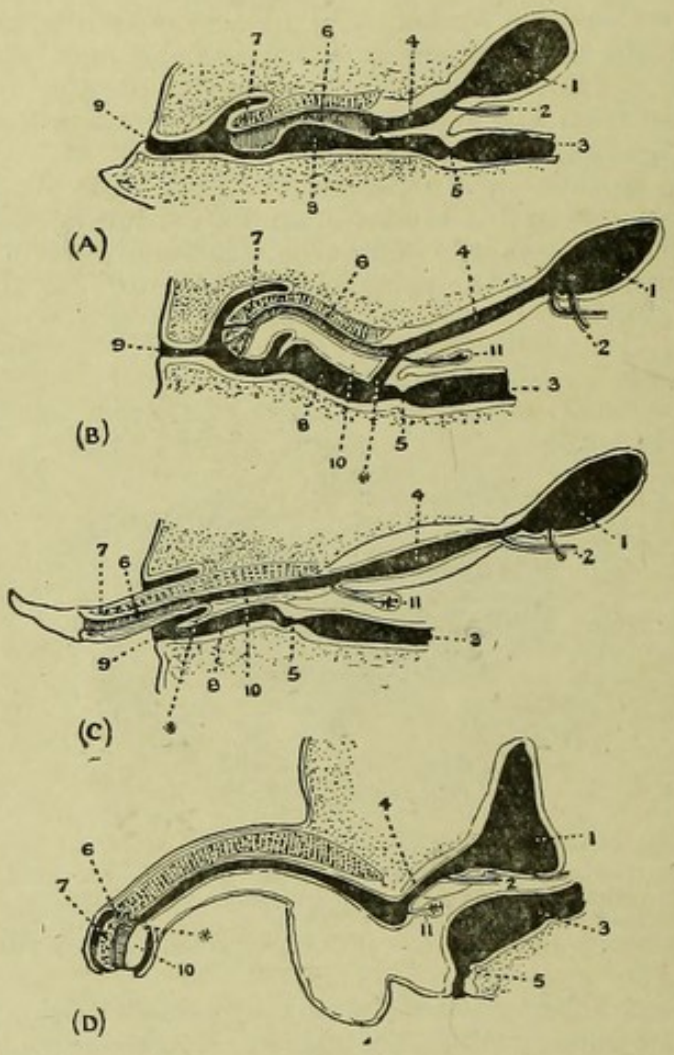

Fig. 10.—Stages in the evolution of the penis. (A) stage seen in tortoise; (B) stage seen in Echidna; (C) stage seen in marsupial (kangaroo); (D) stage seen in man. The phallic canal is shaded. 1, Bladder; 2, Wolffian duct (vas); 3, rectum; 4, urogenital sinus; 5, anus; 6, phallic groove and canal; 7, glans; 8, cloaca; 9, cloacal orifice; 10, floor of phallic canal; 11, Cowper's glands; * position of primitive orifice of urogenital sinus.

canal forms merely a small part of the urethra (see Fig. 10 C). In man (Fig. 10 D) the penis is permanently extracloacal. The urethra is made by the union of the phallic canal and urogenital sinus, but the first element is greatly reduced in extent, forming merely that part of the urethra contained in the glans; were the primitive uro-

genital orifice to remain patent it would be found under the prepuce in the frenum (see Fig. 10 D).

A cursory examination of the appearances seen in the developing perineum of the human fetus supports the history of the penis as just sketched, and helps us to understand the various conditions of hypospadias. During the seventh week (Fig. 11, I) the lips of the cloaca are pushed apart, and the penis, carrying the labia minora and urogenital orifice, rises from the floor of the cloaca and becomes extruded; in the next stage, about a week later (Fig. 11, II) the urogenital orifice is separated from the

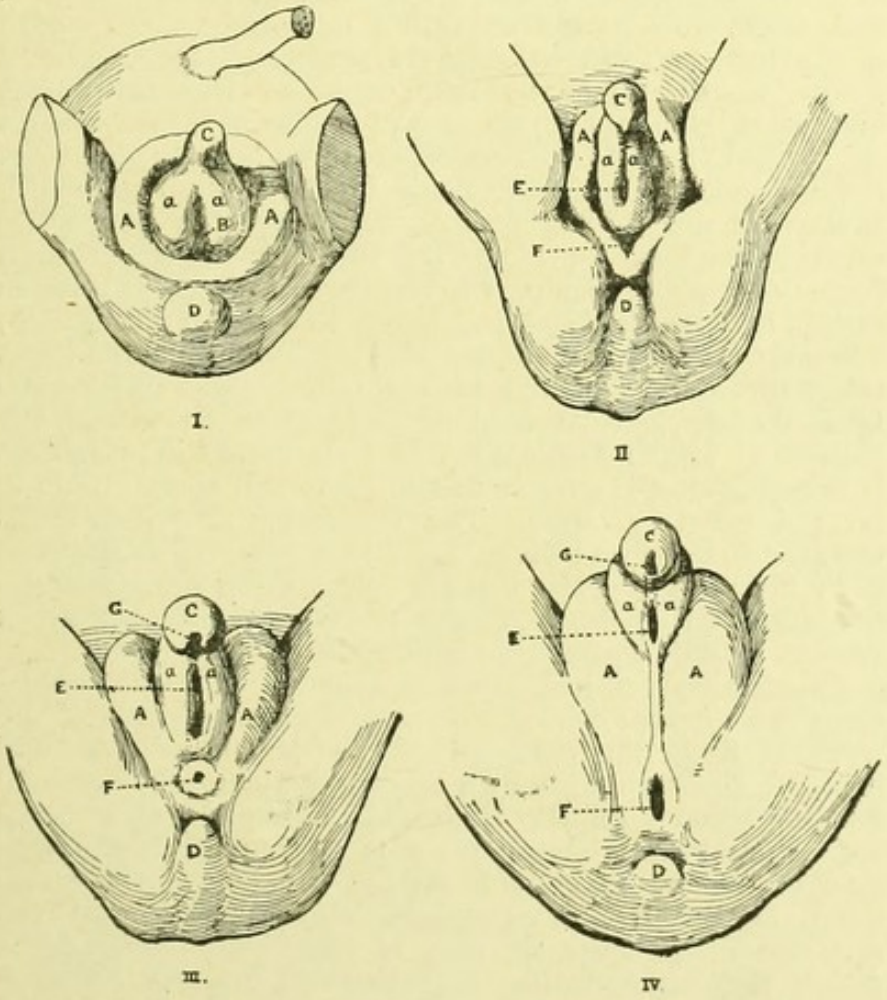

Fig. 11.—Stages in the development of the human penis and perineum. (Drawings by Dr. Stanley Beale after figures given by Kollmann, Keibel, and Hertzog.) I, human embryo 25 mm. long (about seventh week); II, 29 mm. long (about eighth week); III, 31 mm. long (about ninth week); IV, 45 mm. long (about tenth week). A, lips of cloaca (labia majora); B, urogenital orifice being carried to the surface between labia minora (*a a*); C, penis becoming extracloacal; D, tail; E, urogenital orifice; F, anus; G, meatus.

anal orifice by the fusion and growth of the labia minora; the passage for semen and urine is seen being carried forwards with the outgrowing penis, leaving the rectal orifice within the posterior fornix of the cloacal orifice. It is the stage seen in Edentates. In the next stage (Fig. 11, III)

three further changes are seen: (1) The phallic groove on the perineal surface of the glans is being converted into a canal by the union of the lips of the groove; (2) the anal orifice is now apparent on the perineum, and the cloacal lips (labia majora) are fusing together between the anal and urogenital orifices (Fig 11, III); (3) the urogenital orifice, bounded by the labia minora, is being carried out from between the cloacal lips (labia majora) and has assumed a linear form. In a further stage (Fig. 11, IV) the male urethra is almost complete. The orifice of the urogenital sinus has been carried forwards with the outgrowth of the penis, and is now a small opening on the under surface of the penis behind the glans. The phallic canal is formed, and when the urogenital orifice is closed by the fusion of its lips to form the frenum praeputii the urogenital products will escape by that canal at the permanent meatus in the glans. The scrotal raphe is now apparent (Fig. 11, IV), and extends from the anus to the labia minora, which end at the penis in the frenum praeputii. Stages III and IV make clear the manner in which the raphe is formed. In Stage III the floor of the urethra, from the membranous part to the part in the glans (phallic canal), is open, that opening being the urogenital orifice; the labia minora, which form its margin, unite behind in a falciform lamella which is the fourchette. When the labia majora (cloacal lips) unite they do so within the substance of the fourchette; as they unite the fourchette is raised up to the surface of the perineum and scrotum to form a seam or raphe. The fourchette grows forwards towards the glans as the labia majora fuse within it, forming a floor to the penile urethra and uniting the lips of the urogenital orifice (labia minora). Hypospadias represents various stages of arrest of the process just described.

The description just given is founded on the investigations of a number of men. First and foremost there is the work of Keibel[13]; it was he that showed that the seminal canal of Echidna arose by a closure of the phallic groove; Broek[14] has traced the stages in marsupials, and shown that the urethra of the male is a composite structure arising by the fusion of phallic canal and urogenital sinus. Berry Hart[15] recognized at an early date the separate formation of the part of the urethra within the glans, here named the phallic canal; recently Lichtenberg[16] has investigated its formation in man, and pointed out its relationship to the various forms of hypospadias. Fleischmann[17] does not think the margins of the urogenital sinus fuse to form the urethral floor; he regards the urogenital orifice as being carried forwards by the growth of the penis, the floor of the urethra being formed by a corresponding growth of the fourchette.

Having thus given rather a lengthy account of the development and morphology of the external genital organs, I now turn to the preparations illustrating their malformation. The specimens at our disposal are arranged in the accompanying table in seven groups, the classification adopted seeming to me the most natural.

List of Specimens showing Hypospadias and Allied Conditions.

	Museum of Royal College of Surgeons.	Metropolitan School Museums.	Total.
GROUP A:			
Eunuchs	3	0	3
GROUP B:			
Minor degrees of hypospadias ...	1	2	3
GROUP C (Gynaecoid males):			
Males with arrested development of the floor of penis—			
(*a*) Human	4	5	9
(*b*) Animal	13	5	18
GROUP D (Android females):			
Females with development of floor to vulva—			
(*a*) Human	6	10	16
(*b*) Animal	1	0	1
GROUP E:			
Imperforate penile urethra ...	2	3	5
GROUP F:			
Anomalous cases	1	1	2
GROUP G:			
Imperforate vagina	2	4	6
	33	30	63

GROUP A.—CONDITION OF THE PENIS IN EUNUCHS.

In the museum there are three specimens of eunuchs, one belonging to the Barnard Davis collection, another presented by Mr. Edmund Owen, and a third by Dr. Lunn. These specimens are produced now (1) because they show a partial atrophy of all the parts relating to the penis and penile urethra, and (2) because they afford me the opportunity of drawing attention to the very clear conception Hunter had formed of the functional significance of the "bulb of the urethra." The bulb, as Hunter pointed out, is small or almost absent in eunuchs. In most modern textbooks of anatomy the term "bulb" has come to be applied to the enlarged posterior end of the corpus spongiosum, but that was not Hunter's conception; he applies the term to the dilatation of the urethra into which the glands of Cowper pour their secretion, and in which the seminal and prostate fluids accumulate during copulation; the bulb is emptied during the paroxysm by repeated contractions of the bulbo-cavernosus (ejaculator seminis of Hunter—formerly, as a constrictor of the cloaca, it also served this function). The constrictor urethrae, situated on

the proximal side of the bulb, prevents the reflux of its contents towards the bladder. Hunter regarded the bulb and its musculature as an apparatus for the forcible emission of semen; the transformation which is seen to take place in the perineum of the male fetus during the second and third months is connected with the adaptation of the urogenital sinus to serve as an ejaculating and conducting apparatus in place of the cloaca, as in the tortoise. An atrophy of the bulb, of its masculature, or an arrest of development may be regarded as due to the absence or the imperfection of the genital glands.

Group B.—Minor Conditions of Hypospadias.

In my opening demonstration I remarked we should discover both the merits and defects of the museum, and here we reach a very considerable blank, for there is only one preparation in the museum to show the minor degrecs of hypospadias, apparently every one supposing that such a common condition must be already amply represented. In the museums of the metropolitan medical schools I saw only two specimens, and yet in a recent paper Dr. Edington[18] was able to give an account of 28 cases. Thanks to the courtesy of the staff of the London Hospital, I had opportunities of seeing many cases and of forming a first-hand opinion as to the nature of the defects. Minor degrees of hypospadias may be grouped in the three stages shown in Fig. 12; A, stage 1 of that

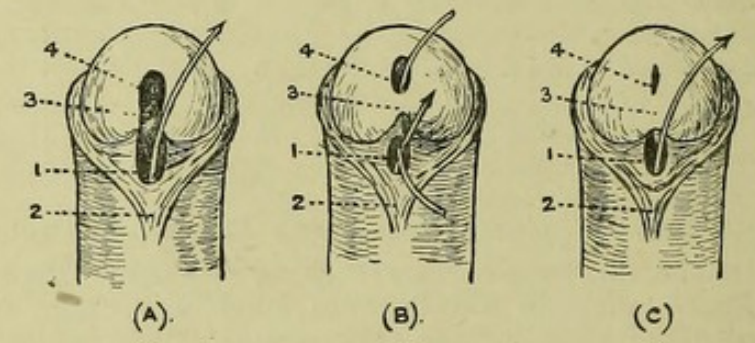

Fig 12.—Three types of hypospadias—(A) in which the groove in the glans (phallic groove) is open, and urine passes by the urogenital orifice; (B) in which the floor of the phallic groove is formed, but the urogenital orifice is unclosed; (C) in which the phallic groove is unformed or obliterated and the urogenital orifice serves as a meatus. (Drawn by W. Finerty.) 1, Urogenital orifice; 2, frenum praeputii; 3, phallic groove and canal; 4, permanent meatus.

figure, shows a condition recalling the tortoise form—the part of the urethra formed by the union of the lips of the glans (phallic groove) remains open, thus forming a furrow which terminates behind at the opening of the urogenital sinus. The orifice of this sinus serves as a meatus; it is bounded by two lateral folds, which are continued forwards into the glans and frenum of the prepuce, while behind the orifice they unite in a sharp, thin falciform margin, which undoubtedly represents the fourchette of the female vulva, and therefore may be

justly named the fourchette. The primitive urethral orifice is thus bounded behind by the fourchette; each side is bounded by a lateral lip continuous behind with the fourchette, and in front with the frenum of the prepuce; and these lateral lips are derivatives of the inner genital folds (labia minora). In Stage II a condition seen in Echidna is represented; the groove in the glans is converted into a canal by the fusion of the lips; but there is still a deficiency between the two parts of the urethra; during micturition most of the urine escapes at this deficiency—the urogenital orifice, but some passes into the canal in the glans (phallic canal) and escapes by the permanent meatus.

Stage III represents a condition which is intermediate between the two just described. The groove on the glans is at first filled with epithelium, which serves as a bridge or medium for the fusion of the mesoblast or connective tissue in the lips of the glans. When the fusion of the lips has taken place desquamation of the epithelium thus enclosed in the glans occurs and a canal is formed. If desquamation fails, then the form shown by Stage III is the result. Dr. Berry Hart, who has done so much to elucidate the development of the genital organs, regards the formation of the canal in the glans as a result of the ingrowth of a rod of epithelium, but an appeal to comparative anatomy and on the embryological evidence placed before me by Dr. J. E. Spicer, I have no doubt that the interpretation given here is the more accurate.

In most cases of hypospadias there are seen on the dorsal surface of the prepuce two groups of sebaceous glands forming raised brownish oval areas; they represent the preputial glands of other mammals, and are found constantly in the female in the recess behind the prepuce of the clitoris. They have been described at length by Mr. Shillitoe and myself.[19]

The closure of the urogenital orifice (see Figs. 11 and 12), and the complete fusion of the urogenital sinus with the phallic part of the urethra takes place by a fusion of the fourchette and lateral margins of the urogenital orifice in the frenum of the prepuce (Fig. 12). Thus the minor degrees of hypospadias represent arrested stages of development, and it is probable that the arrest will be found, as Hunter suggested, in some disturbance in the later development of the testes in the fetus. In most cases of minor hypospadias the sexual functions are perfect in the adult.

Group C.—Males with Arrest of Growth of the Floor of the Penis (Gynaecoid Males).

In this group the testes are permanently imperfect, whereas in the last group the testes, at least in the child and adult, appear to yield the secretions necessary for the growth and full emotional life of the individual. The condition of the external organs seen in this group of cases often represents quite early stages of the development of the penis. In the College museum there

are 17 specimens, 4 of which are from the human subject; in the metropolitan medical museums there are 10, 5 of which are human. The specimen represented in Fig. 13 may be taken as typical of the group. The penis is only partly extruded between the lips of the cloaca (labia majora) which form the scrotum; it has been arrested in the process of becoming an intracloacal organ. The arrest in the development of the bulb and all the apparatus necessary for the ejaculation of semen is still

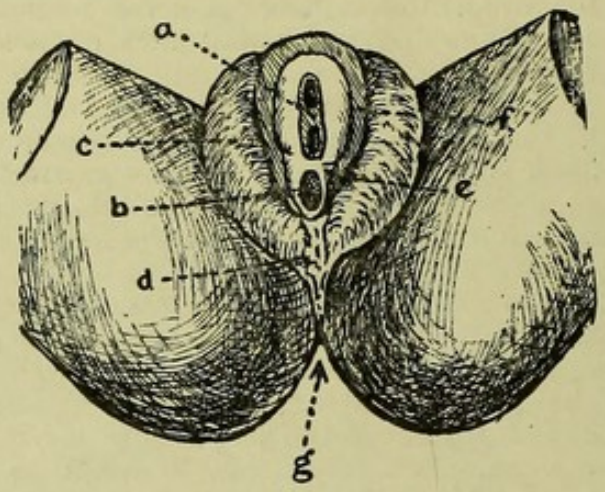

Fig. 13.—Hypospadias in a gynaecoid male (perineal view). (Drawn by William Finerty.) *a*, Phallic groove on under surface of glans; the posterior depression in the groove represents the lacuna magna; *b*, urogenital orifice; *c*, labium majus, uniting behind with opposite lip to form scrotum, *d*; *e*, labium minus; *f*, prepuce; *g*, anus.

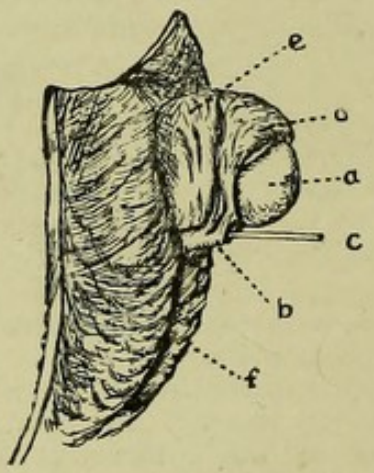

Fig. 14.—Similar specimen to last, seen in profile. *a*, Glans, *b*, fourchette; *c*, urogenital orifice; *d*, prepuce; *e*, skin of body of penis; *f*, scrotum.

more marked. The glans is merely grooved; the meatus shown is the orifice of the urogenital sinus; it is bounded by the fourchette behind, and on each side by folds representing the labia minora.

The lateral lips of the orifice end in front on the glans and also in the prepuce on each side of the glans; behind they are continuous with the fourchette. In the fourchette

two other lateral folds are seen to meet and fuse, the point of fusion giving rise to the median raphe. The two folds on each side are (1) the labia majora (the cloacal lips); (2) the intermediate folds, which seem to have escaped notice by former observers. These folds, when traced forwards, become continuous with the skin of the penis—not with the prepuce or skin of the glans, which is an old structure, but with the skin of the body of the penis, which is, comparatively speaking, a new structure. The intermediate folds also meet behind within the fourchette. The fourchette is the primary scaffolding in which the labia majora and intermediate folds of the two sides fuse, and as they fuse the substance of the fourchette is raised up to form the raphe. The degree of fusion varies greatly; it may be little more than in the female fetus, or it may amount to an almost complete fusion of the scrotal halves. One specimen in the museum shows an extreme degree of arrest, the urogenital and anal orifices being minute in size, and grouped close together in a small flat field on the perineum. In many of these cases the vagina, uterus, and tubes are less atrophied than they are in the normal male child. It seems possible that the internal secretion of the testes causes not only a growth of the parts which are developed in the male, but also an atrophy of the parts which are characteristic of the female. The internal glands may be testicular in structure yet the external organs may be exactly of the female type.

Group D.—Android Females.

Females Showing the Development of a Floor to the Vulva. (Hyposynclesis.)*

Six specimens in the College museum illustrate this condition (Fig. 7, Demonstration I), while there are ten in the museums of the metropolitan medical schools. In all of these cases the sexual gland simulates an ovary; the vagina at its orifice is always constricted; the prostate is often developed to a considerable degree; the Wolffian ducts are retained to a greater degree than in the normal female. The extent to which hyposynclesis (the union of the labia to form the penile urethra and scrotum) takes place varies in extent; Fig. 15 represents a specimen in the museum of St. George's Hospital Medical School, which shows the process carried to a minor degree; the falciform web in which the labia majora terminate behind represents a greatly developed fourchette; Macnaughton-Jones[20] has recently reported such a case and has seen another. Great hypertrophy of the fourchette is accompanied, I believe, with an incomplete development of the internal genital organs. Blacker[21] has shown that in

* A word to designate the opposite condition to hypospadias is necessary; Mr. Plarr, Librarian to the College, has given me the term employed above.

15 per cent. of women the fourchette is continuous with the labia minora and in 85 per cent. with the labia majora. In the common type of android females—individuals with the internal organs of the female type and the external simulating more the male type than the female—the condition of the penis and scrotum is exactly similar to that seen in gynaecoid males (see Figs. 13, 14). The condition of the external genitals gives no clue to the sex of the internal, but one may justly infer that, whether they are ovaries or testicles, they are in either case imperfect and non-functional. In one case I found that what was unmistakably an ovary from outward appearance was on microscopical examination made up chiefly of tubules which were embedded in a richly nucleated cellular stroma. It is usually said that in the normal female the external genital organs represent the condition of the young embryo, and that positive changes only occur in the male. This appears not to be

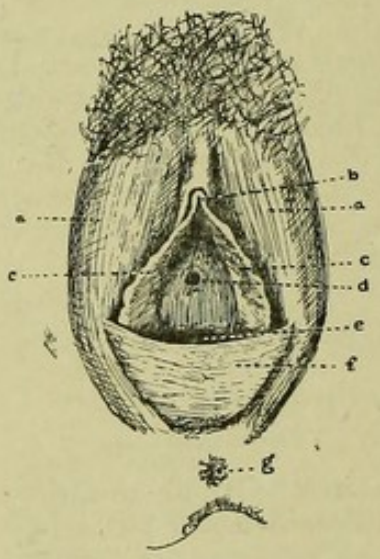

Fig. 15.—From a specimen in the museum of the medical school of St. George's Hospital. *a*, Labia majora; *b*, clitoris; *c c*, labia minora; *d*, urethra; *e*, orifice of vagina; *f*, enlarged fourchette; *g*, anus.

the case; while in the male the urogenital orifice becomes closed to form the urethra, in the female it opens up to form the vulva. The process in either case is evidently determined by the internal secretion of the sex gland, so that a neutral condition of the external sex organs is an indication of a similar condition in the internal glands. In many primates, especially South American monkeys, the clitoris in the female is nearly the size of the penis in the male, but is furnished with a groove in place of a canal.

Group E.—Imperforate Penile Urethra.

Occasionally cases occur of congenital occlusion of the urethra within the glans, but of such we have no example in the College collection. Occlusion most frequently

occurs between the triangular ligament and the glans, and often the whole of this tract is affected. There are but two specimens in the College museum, and three in other London museums. One of the two College specimens was very thoroughly examined by Dr. J. E. Spicer,[22] and presented by him to the museum; his preparations showed an irregular core of epithelium within the corpus spongiosum representing the urethra from the membranous part almost to the glans. The bladder, prostatic and membranous parts of the urethra were so distended that the child, although only in the sixth month of development, was of such circumference that it was delivered with great difficulty. The part of the urethra thus imperfectly developed represents the part which Hunter regarded as specially adapted for the ejaculation of semen. We have seen that the floor of this part of the urethra is formed by the growth of the fourchette. Now, the growing edge of the fourchette is embedded in a mass of epithelium which fills the primitive urogenital orifice as that orifice is carried forwards. Thus the whole of the penile urethra is at first represented by a solid rod of epithelium, which afterwards becomes canaliculized by a process of desquamation from within. These cases represent an arrest of the process of canaliculization, and probably are due to an imperfection of the testicles. In Dr. Spicer's case the rectum opened in the urethra.

Group F.—Anomalous Cases.

In this group there are only two cases. In one of these there is a complete arrest of the external genital organs; No. 703 is the pelvic viscera of a girl of 7 years, presented to the museum collection by Mr. Page. In it there are three openings on the perineum: (1) the anus, (2) the vagina, (3) the urethra opening at the tip of an enlarged clitoris. The specimen is interesting, because a condition somewhat similar is met with in lemurs and allied animals. In these forms the urethra in the female, instead of opening into the vulval cleft, is carried forwards on to the clitoris, and thus opens outside the vulval cleft.

Group G.—Imperforate Conditions of the Vagina.

So far all the specimens dealt with—imperforate anus, hypospadias in the male, and hyposynclesis in the female—all pertain to the transformation of the cloaca into the sexual organ of the male. But there is also, especially in the human body, a sexual transformation of the cloaca in the female. Dr. Berry Hart and Dr. Wood Jones have investigated these changes, which they found to occur in the female human fetus from the tenth to the thirteenth week (fetuses measuring 60 mm. to 140 mm.). It has already been shown that after the migration of the rectum to the perineum, the endodermal cloaca becomes the trigone of the bladder and the urogenital sinus (see Figs. 3 and 4 of first demonstration). In the early fetus (see Fig. 16)

the sinus is a long tube receiving high up the genital ducts and urethra. About the commencement of the tenth week the epithelium lining the terminal parts of the Müllerian ducts proliferates (Berry Hart believes the proliferation takes place from the Wolffian ducts), and grows towards the perineum as two solid rods; the mesoblast surrounding the rods is also in a state of active growth. The rods fuse together, the hymen and lower third of the vagina being formed by a canaliculization of the epithelial rods. The transformation of the terminal parts of the Müllerian ducts results in the opening of the vagina being brought down almost to the perineum; all that remains in the female of the original cloaca are the trigone of the bladder, the urethra, and that

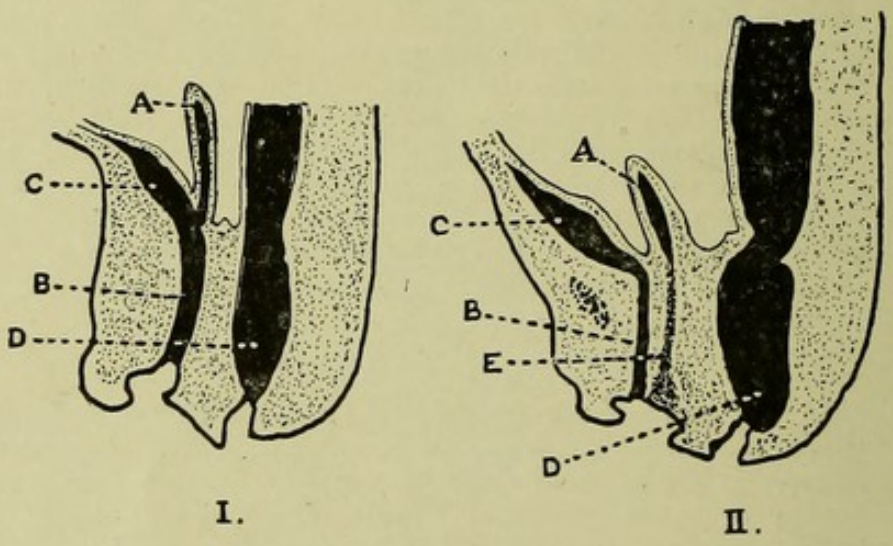

Fig. 16.—Diagrams showing the termination of the vagina about the seventh week (I) and about the thirteenth week (II); after Dr. F. Wood Jones. A, Müllerian ducts (vagina and uterus); B, urogenital sinus: C, bladder; D, rectum; E, vagina represented by a rod of epithelium.

part of the vulval cleft which lies between the labia minora. The migration of the vaginal orifice is a sexual change—an adaptation for impregnation and parturition. In all the cases of android females—females in which hyposynclesis had occurred—two things were noticed about the vagina: (1) the orifice was high up in the urogenital sinus, a position which is normally retained in the male; (2) it is very narrow—clearly in these cases the vaginal migration had failed to occur. In three of the six android females the rectum opened into the clitoric urethra.

Thus it will be seen that Hunter's dictum, that the full development of the sexual characters depends on the testicular or ovarian stimulus of perfection, seems likely to provide the best key to the pathology of all the cases we have considered, and that a more exact study of such malformations is likely to throw much light on certain obscure physiological problems.

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III.—ECTOPIA VESICAE AND ALLIED CONDITIONS.

For the purpose of this demonstration it is convenient to divide the specimens at our disposal into four groups: (1) Ectopia vesicae in the male; (2) ectopia vesicae in the female; (3) ectopia vesicae combined with fistulous condition of the intestine; (4) epispadias.

Group I.—Ectopia Vesicae in the Male.

There are 3 specimens of this condition in the College museum and 16 in the museums of the metropolitan schools, making 19 altogether. Of the 19, 10 are from adults; the others are from infants, ranging in age from birth up to 3 years. All the specimens show the same lesion; the description of the one shown in Fig. 17 may serve for the rest. The mucous membrane of the bladder and urethra, from urachus above to meatus below, is spread out on the surface of the hypogastrium, and becomes continuous with the skin of the belly wall at its margin. Four areas may be recognized in the exposed mucous surface—especially in specimens where the vessels and capillaries have been successfully injected. (1) The upper area, crescentic in shape, is really skin in which the superficial layer of epithelium has not undergone a corneous change. This crescentic area may be named the hypogastric fold, for, like the fourchette of the vulva, it serves as a bridge for the fusion of the embryonic tissues out of which are formed muscular, bony and connective structures in the mid-line of the body wall—in a manner to be described later. (2) Below the hypogastric fold is a rough villous area of vesical mucous membrane, which may be distinguished as the upper vesical area; (3) an inferior vesical area, crescentic in shape and differing from the upper area in texture; it represents the trigone of the bladder and prostatic part of the urethra, and is a derivative of the urogenital sinus. On it open the ureters, uterus masculinus and common ejaculatory ducts (Fig. 17). (4) An area on the upper surface of the penis representing the spongy part of the urethra. The prepuce forms a crescentic fold below the glans; the frenum, the under surface of the penis and the

scrotum, are normally formed. On each side of the inferior vesical area is seen an upward continuation of the scrotal folds (labia majora).

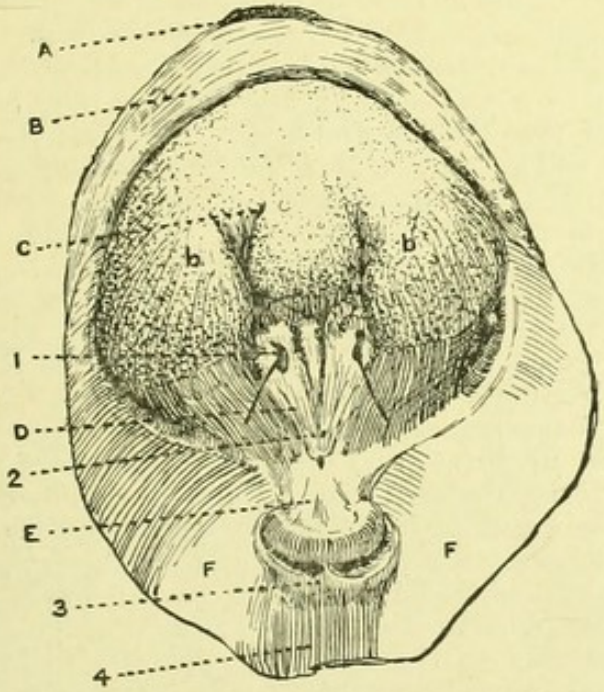

Fig. 17.—Ectopia vesicae in male (Dr. Stanley Beale). A, Position of umbilicus; B, hypogastric fold; C, upper vesical area; D, lower vesical area, with openings of ureters (1) and common ejaculatory ducts (2); E, penile or spongy area; F, continuation of labia majora; 3, prepuce below glans; 4, scrotum.

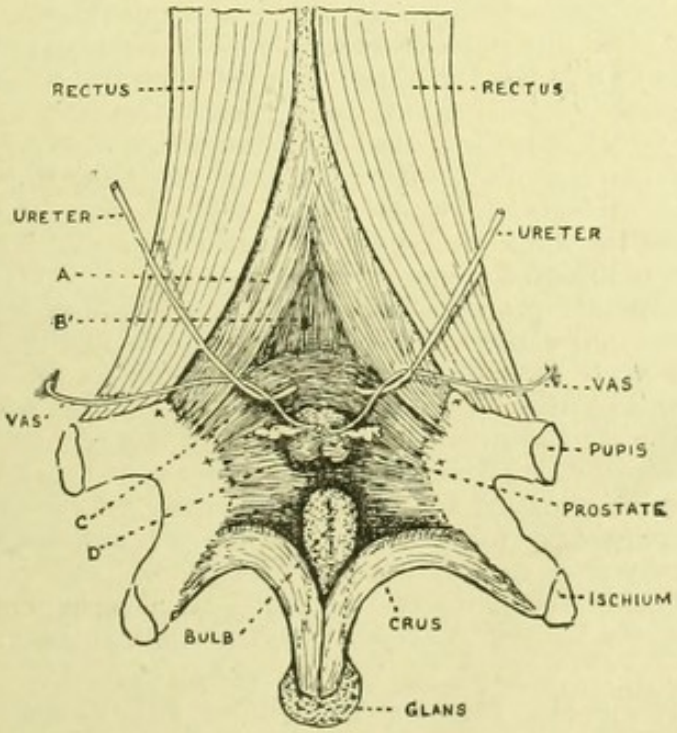

Fig. 18.—Bladder in ectopia seen from its pelvic aspect. A, Vertical pubo-vesical fibres; B, vertical or detrusor fibres; C, oblique and horizontal pubo-vesical fibres; D, muscular fibres representing constrictor urethrae and deep transverse perineal muscle; XX, margin of pubis which normally enters into formation of symphysis.

Unfortunately, the College collection is poor in specimens illustrating the anatomy of the deeper parts in ectopia vesicae; indeed, with the exception of a paper by Professor Shattock,[23] there is little to be found in medical literature concerning this subject. From specimens in the museums of St. Thomas's, Guy's, and Middlesex Hospitals, I have been able to construct Fig. 18, which illustrates the connexions of the bladder and penis when these parts are dissected and viewed on their pelvic aspect. The bladder wall fills the triangular gap which separates the one rectus from the other just above the widely separated pubic bones. As Professor Shattock has shown, nearly all the musculature of the bladder in cases of ectopia arises from the posterior aspect of the pubes : (1) Fibres arise from the descending pubic rami and pass to the trigone of the bladder, to the prostate, and to the bulb, many of these representing the deep transverse perinei, constrictor urethrae and pubo-prostatic fibres of the normally formed males; rising from the pubis above these are (2) fibres passing transversely on to the bladder and blending with similar fibres rising from the opposite side; (3) vertical fibres which ascend from the pubis to end in the bladder and in the fibrous tissue which binds the vesical margins to the adjacent edges of the two recti. The vesical musculature, having to serve as part of the abdominal wall, and being subject to all the strains that fall on that wall, becomes greatly hypertrophied and evaginated, and forms a hernial sac. Professor Shattock is probably right also in attributing this hypertrophy in part to the strain which falls on the muscular tissues which serve to bind the pubic bones together; a strain must fall here, for while at birth the gap between the bones measures about 1 in. in adults, the separation usually amounts to 3 or 4 in. It is clear that if an operation is to be undertaken with the view of forming a functional bladder, it must be undertaken at the very earliest date, otherwise the organ becomes irremediably malformed and incapable of being moulded to form a hollow sac.

The prostate projects backwards from the fibro-muscular tissue spanning the symphysis; it is usually bilobed and covered with muscular tissue; the common ejaculatory ducts receive the vasa deferentia and vesiculae seminales and perforate the prostate. The bulb of the corpus spongiosum is well formed, and as it passes forwards (see Fig. 18) comes to lie, not on the under but on the upper surface of the corpora cavernosa, which are shorter and united less completely than in the normal condition. The reversion in position of the parts of the penis must be kept in mind in shaping a theory to account for the malformation.

Group II.—Ectopia Vesicae in the Female.

This group is represented in the College collection by a pelvis, and the cast of the adult woman of whom the pelvis was part, the specimen having been presented early last century by Professor Mayo. In the museums of the

metropolitan medical schools there are six specimens, all obtained from infants, with the exception of No. 2695 in the museum of St. Thomas's Hospital School, which was prepared from a woman aged 22 years. Lately Mr. Lawford Knaggs has described a case [24] in a woman aged 31, from whom he removed a normally-formed uterus and implanted the ureters in the rectum. In four of the six museum specimens the uterus and vagina are normally developed; in the remaining two there was a division of the uterus and vagina.

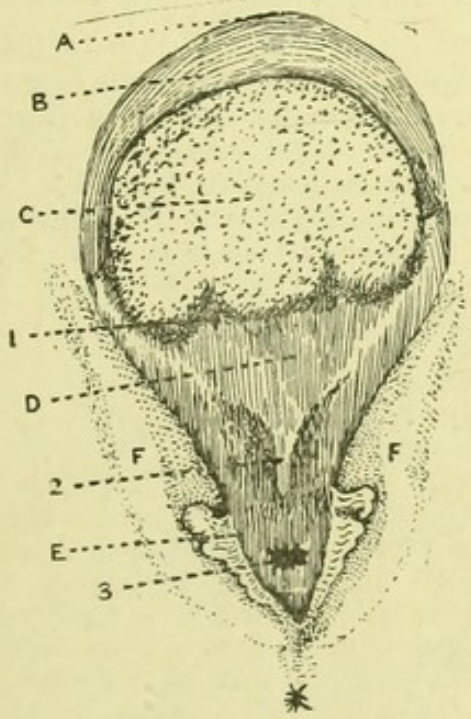

Fig. 19.—Ectopia vesicae in the female. A, Position of umbilicus; B, hypogastric fold; C, upper vesical area; D, lower vesical area; E, labium minus with half of clitoris and prepuce: F, continuation of labia majora. 1, Opening of the ureter; 2, uvula vesicae; 3, vaginal orifice.

The condition of parts in ectopia in the female is shown in Fig. 19. The red exposed mucous surface commences below just anterior to the anus as a sharp angular recess between the lower extremities of the labia majora; on the floor of this recess is seen the orifice of the vagina; on each side of the vaginal orifice is a process representing a labium minus, with the corresponding crus and half of the glans and prepuce. On a smooth part of the exposed mucous surface (the lower vesical area) are seen the orifices of the ureters and the uvula vesicae; the mucous membrane of the upper vesical area is rough, villous, and corrugated, and often shows a symmetrical division into right and left halves, due to the traction of the ureters on the evaginated bladder. In a specimen of Dr. Champneys's in St. Bartholomew's Hospital Museum with double vagina and uterus, there is an opening on the mucous area behind the vaginal orifices leading into a cul-de-sac of

uncertain nature. As in the male the upper part of the exposed mucous area is formed by the hypogastric fold.

It will be observed that in the female the clitoris is cleft, the labia majora and minora are separated, the vulval cleft being added to the widely cleft urethra and bladder, whereas in the male, the parts representing the margins of the vulval cleft and clitoris were united. Hyposynclesis being a secondary and later process than the production of ectopia, is not affected.

Group III.—Ectopia Vesicae with a Meckel's Diverticulum opening on the exposed Vesical Surface.

There are eight specimens in the museum illustrating this condition, five from females and three from males; in the museums of metropolitan medical schools there are six preparations (four males, two females). The condition appears to occur as frequently in males as in females, whereas pure ectopia vesicae is three to four times more

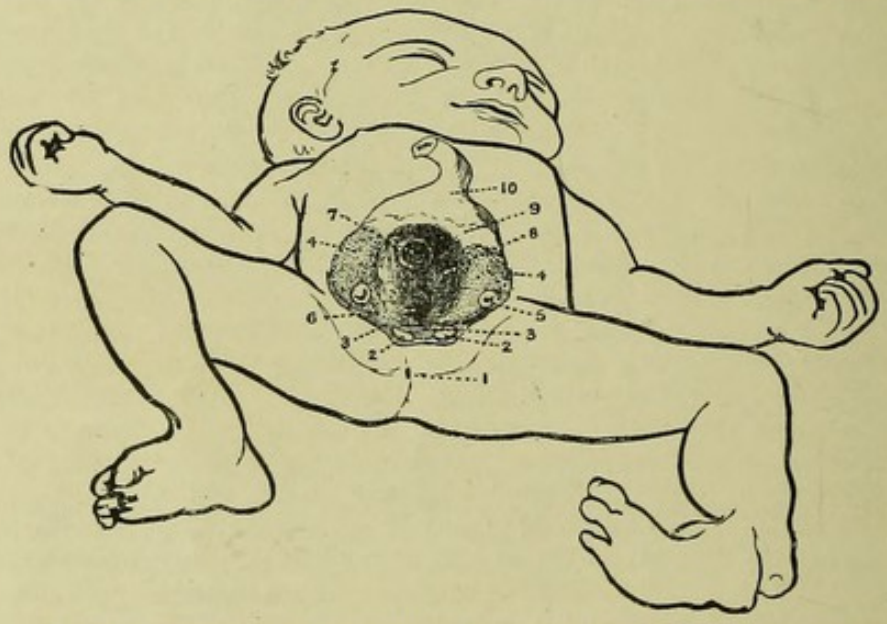

Fig. 20.—Ectopia vesicae with inclusion of part of a yolk sac in vesical mucous membrane (modified from Bockenheimer, by Dr. Stanley Beale). 1, Anus; 2, labium minus with half of clitoris; 3, lower vesical area; 4, upper vesical area; 5, opening of Müllerian duct, opening of ureter immediately below; 6, recess leading to caecum and colon; 7, recess leading to ileum; 8, enteric or vitelline depression between halves of bladder; 9, hypogastric fold; 10, umbilical cord.

frequent in males than in females. Recently Professor A. M. Paterson and Dr. Emrys-Roberts[25] gave a very full description of a case, while Mr. B. Ley and I had an opportunity given to us by Dr. James Greenwood of dissecting such a specimen. In all of these cases the child was either born dead, often at the seventh month of pregnancy, or died a day or two after birth. The importance of the condition is that it seems to give a clue to the manner in which ectopia vesicae may be produced. The condition is so alike in all the specimens examined that the description of one serves for the rest.

Fig. 20 shows the condition of parts on the ventral aspect, from umbilicus to perineum, in a female child in which this deformity was present. Near the middle of the exposed area of mucous membrane there is a depression; into the upper part of this depression opens the ileum; into the lower part opens the caecum, together with a diverticulum representing the colon and rectum. The depression into which the ileum and caecum open occupies the position of a Meckel's diverticulum, and undoubtedly represents a part of the yolk sac. The depression is usually lined by a villous mucous membrane. The median depression has on each side of it an area of vesical mucous membrane, the two areas representing a divided upper vesical area of the usual type of ectopic bladder. At the margin of the vitelline depression the intestinal mucous membrane becomes continuous with the vesical and hypogastric zones. On the smooth mucous area (inferior vesical area) below each half of the bladder open (1) the Müllerian duct, for in none of these cases have the Müllerian ducts fused to form a single vagina and uterus; (2) below the Müllerian or vaginal orifice, the ureter (Wolffian duct). The vulval cleft is widely open, the halves of the clitoris, prepuce, and labia minora being represented by tags on the margins of the exposed mucous area (see Fig. 20). In the male there is a corresponding condition, the halves of the penis and scrotum being ununited. The exposed mucous surface usually reaches up as far as the umbilicus, and the viscera of the abdomen are always extruded to a greater or less degree at the umbilicus and attachment of the cord. The intestinal or vitelline orifice varies in position; it may be situated almost in the perineum near the posterior angle of the exposed mucous area. In half of the cases examined the small intestine had become prolapsed through the vitelline orifice. The rectum is imperforate and usually forms merely a caecal process, which may be hard to distinguish from the caecum proper; the appendix is occasionally bifid; a proctodaeum is usually present. Spina bifida is present without exception. The orifices of the Müllerian ducts may be occluded. The ureter may open into the Wolffian duct, or separately on the mucous surface of the crescentic-shaped trigone. The testicles are undescended in the males. In the College specimen No. 668, so well described in the *Journal of Anatomy*, vol. xvi, 1881, p. 226, by Mr. Alban Doran, a narrow tube, with an epidermal lining, passes from the proctodaeum to open in the lower vesical area.

Group IV.—Epispadias.

There is one specimen illustrating this condition in the College collection, and two in the museums of the metropolitan medical schools. The condition differs from ectopia in degree only. Instead of the four exposed areas to be recognized in ectopia, there is here only one of them

—the spongy area—but by pressing aside the loose curtain-like fold above the penis and distending the wide folded passage which serves as a urethra, the lower vesical area can also be seen. On the other hand, the upper vesical area has been closed over in the normal manner, and thus the upper part of the bladder is complete; the symphysis is normal, while the hypogastric fold forms the loose curtain which bounds the upper border of the epispadiac urethra. Cases are seen, but we have no specimen in the College collection, where the upper vesical area remains open, while the lower vesical area and spongy area are folded in in a perfectly normal manner.

Nature of Ectopia Vesicae.

It seemed possible that the cases where ectopia vesicae is combined with a fistulous condition of the intestine might provide a clue to the abnormal developmental processes which culminate in the obscure condition known as extroversion, or ectopia of the bladder. There can be no doubt, in the third group of cases here described, that the yolk sac, or part of the yolk sac, opens on the surface of the body and there becomes continuous with the vesical mucous membrane. To find the key to the condition, one must return to that stage of development when the rudiment of the bladder is continuous with the yolk sac. That is best shown in Graf Spee's human embryo, which is probably in the third week of development. (See Fig. 21.) It shows a process, continuous with the yolk sac, growing into the body stalk. There is no doubt as to what that process is: it is the allantois, but unfortunately the allantois is a structure which is little more than a name to most of us. Now it is certain that in the evolution of the vertebrates a bladder appeared long before there was an allantois, and we have seen that the bladder arises developmentally as a diverticulum from the most posterior part of the hind gut—the endodermal cloaca. The hind gut is an outgrowth from the archenteron—the main part of which remains as the yolk sac. Thus at one point of development, the bladder and yolk sac are in continuity. In the course of the evolution of the higher vertebrates the fundus of the bladder becomes modified to serve the temporary purposes of the embryo. By expanding and forming a connexion with the chorion, the modified fundus of the bladder or allantois serves the purpose of a lung to the embryo. Such a modification is easily conceivable, when it is remembered that the yolk sac itself originally served that purpose, and does so even in the human embryo for a short time, and that the vesical rudiment is originally continuous with the yolk sac. Although the bladder appears much later than all the other parts of the alimentary canal in the evolution of the mammalian body, yet, owing to the important part it plays in the life of the embryo, its appearance is antedated during development, so that it is separated from the archenteron before any other part of the alimentary system. That is what is seen

to be taking place in the Graf Spee embryo. In order to obtain the condition of parts where the vesical mucous membrane is continuous with the bowel through the yolk sac, one must conceive that the rudiment of the allantois (bladder) has not been separated from the yolk sac; that at the period when its separation was due some process was at work which arrested its separation. To see if there was any real foundation for such a guess, I turned to the splendid investigations which Professor Mall[26] has recently published on malformed human embryos (163 in number).

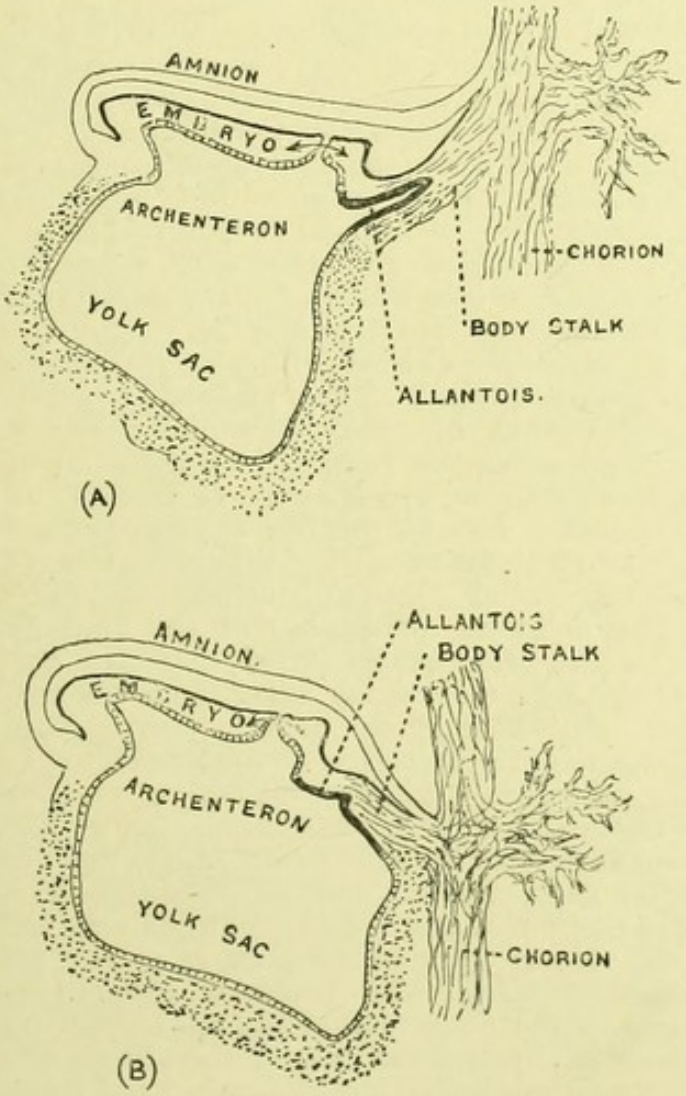

Fig. 21.—(A) Diagram of Graf Spee's human ovum, with the allantoic outgrowth and cloacal region of the archenteron marked in black. (B) The modification which is supposed to proceed ectopia vesicae with fistulous opening of the bowel.

Three of the embryos described by him help us to understand the condition of ectopia. In No. 396 he found that the vessels of the yolk sac grew directly into the chorion, the blood from the placenta being thus returned by the portal vein. I have seen that condition in a full-time child, in which exomphalos was present with ectopia

vesicae. In No. 336 of Mall's series, a human embryo in the fourth week of development, he found the allantoic vessels undergoing obliteration and being replaced by those of the yolk sac. In No. 24 he found the condition I surmised; the allantois grew directly from the yolk sac, and branched as it spread outwards to the chorion. Here the separation of the bladder rudiment from the yolk sac has been arrested by the fusion of the allantois with the yolk sac.

In Fig. 21 (B) is represented the abnormal relations of the hind end of the embryo which results from a non-separation of the allantois from the yolk sac. The hind end of the embryo, with the primitive streak, rests directly on the yolk sac, instead of being separated from it by the allantois and cloaca, as in the Graf Spee embryo (Fig. 21 A). The primitive streak thus comes to rest directly over that part of the yolk sac where the unseparated rudiment of the cloaca is situated. If an opening were to occur in the primitive streak, the yolk sac and endodermal cloaca would open on the hypogastric region of the embryo, and give rise to the condition seen where ectopia vesicae is combined with a fistula opening into the intestine.

What is the primitive streak, and what becomes of it in the normal development of the embryo? All lines of investigation tend to show that the primitive streak represents the opening of the gastrula or body cavity of lower vertebrates, an opening which serves as mouth and anus; in the vertebrate embryo the lips are approximated and drawn out into a linear form—the primitive streak. Along the streak the epithelial covering (ectoderm) of the embryo fuses with the lining (endoderm) of the hind gut. Its position on the adult body may be indicated by a line drawn from the sacral region along the perineum to the umbilicus. The floor of the middle part of the streak breaks down to form the orifice of the endodermal cloaca, and in the woman this primitive opening is represented by the floor of the vulval cleft. The rectum, as we have seen, terminated at first in the endodermal cloaca, but separates to open on the perineum, but the permanent anus is situated in the line of the primitive streak. In front and behind the cloacal opening the margins of the primitive streak become fused; the posterior (caudal) part is obliterated by the outgrowth of the tail end of the body; the anterior or hypogastric part of the streak—from the cloacal orifice to the umbilicus—is invaded and fused by the mesoblast, in which is formed all the tissue in the median ventral line from the skin to the mucous membrane of the urethra and bladder.

It is plain, as Reichel[27] was the first to show (1898), that ectopia vesicae is due to a non-fusion of the lips of the hypogastric stretch of the primitive streak; if the theory of the primitive streak being the coelenterate mouth be absolutely true, we have in ectopia vesicae an arrest of development that restores a large part of that mouth.

What are the most likely conditions to arrest the proper fusion of the lips of the primitive streak? Hare-lip, cleft

palate, anencephaly, and some forms of spina bifida are examples of a similar failure of fusion. Experimental embryologists—especially Loeb—have shown that such failures can be produced by adding solutions of salts to the medium in which the embryo grows; but Mall's observation that the chorion is inflamed—probably a result of endometritis—in practically every case where the embryo is malformed, seems more helpful for our purpose. Chorionic inflammation means a disturbed and at least a temporary derangement of the embryonic circulation. The developmental fusion of processes or folds is akin to the healing of a wound; the tissues at the healing edge being the least resistant undergo dissolution first when the nourishment of the tissues is interrupted. Growth may go on in the main parts, and when the embryonic circulation has been restored the edges intended to unite have moved so far apart that a gap has been formed across which the tissues cannot grow. Such a condition is most likely to occur during the third week of embryonic life, when the chorionic circulation is being established, and that is just when the lips of the primitive streak are in a state of fusion, a fusion which is to secure the anterior closure of the bladder and lower belly wall.

On referring to the literature on ectopia vesicae, I found that Dr. Eugen Enderlen,[28] in examining the histological structure of the bladder in 7 cases of ectopia, had found in every one of them that there were areas in which the structure of the mucous membrane was similar to that of the great intestine. It seemed to me possible that these areas might really be included parts of the yolk sac, seeing that there are numerous cases of ectopia in which the bladder is fused with the yolk sac. In ordinary cases of ectopia vesicae I supposed that the vitelline duct, or stalk of the yolk sac, had been obliterated, as occurs in the normal course of development, and that all connexion between the bowel and a remnant of the yolk sac in the bladder was thereby lost. I found that Professor Shattock[23] had published the results of an investigation into the histology of the ectopic bladder in 1894—ten years before Dr. Enderlen's account appeared. He, too, found that the whole exposed surface of the bladder was the seat of an adenomatous growth—somewhat similar to which is seen in the intestinal mucosa.

Sections were cut of two specimens in the College collection. One was a preparation in which the intestinal lining became continuous with the vesical mucous membrane, a section being made at the junctional region. The other specimen was cut from a case of unmixed ectopia. In the first specimen the part supposed to be yolk sac or its stalk was lined by a double layer of columnar epithelium, while that supposed to be vesical mucous membrane was covered by a stratified epithelium similar to that which covers the lips of the mouth. Sections from the second specimen were taken from various parts of the upper vesical area; unfortunately all the epithelium had been lost, but it showed villous pro-

cesses with depressions between their bases, recalling the structure of the small intestine, and quite unlike the mucous membrane of the normal fetal bladder. The results so far are negative, but the matter deserves further investigation on fresh material. In the meantime I am inclined to regard ectopia vesicae with a fistulous communication as the result of an earlier arrest than the unmixed form of ectopia; that in the first form arrest occurs before the separation of the allantois and cloaca from the yolk sac, while in the unmixed form arrest occurs after the separation, and that both malformations are due to a disturbance of the chorionic circulation caused by endometritis.

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