ANOMALIES OF THE FEMALE GENERATIVE ORGANS.

By anomalies of the female generative organs we mean the congenital (not acquired) partial or total absence, the arrest of, or excessive development, or a peculiar formation or malposition of

Development of the External Genital Organs—diagrammatic. 1. P, rectum, continuous with A, allantois (bladder), and M, Müller's canal (vagina); x, depression of the integument below the median tubercle, which by its progress inward forms the vulva. 2. The depression has extended inward to become continuous with the rectum and the allantois to form the cloaca, C. 3. The cloaca has split into the uro-genital sinus, S, and the anus, u, by the down growth of the perineal septum. The Müllerian canals are fused to form the vagina, V, behind the bladder, B, and the orifice of the urethra, u. 4. The perineum completely formed. 5. The upper portion of the uro-genital sinus contracts to form the urethra; the lower portion persists and forms the vestibule, w, into which both urethra and vagina empty.

Malformation of the External Genital Organs—diagrammatic. 1. Complete atresia of the vulva: r, rectum; g, genital canal; b, bladder, communicating with both. 2. Complete atresia of the vulva: r, rectum, separated from the allantois; b, bladder, and g, genital canal, distended with urine. 3. Atresia of vagina and anus; d, perineum, incomplete; b, bladder; v, vagina, and r, rectum, open by a common cloaca. 4. Hypoplasias in the female: first degree coincident with hypertrophy of the clitoris; s, persistent uro-genital sinus, to which succeeds the long vestibular canal; u, urethra, and v, vagina, opening into the vestibular canal; c, hypertrophied clitoris. 5. Hypoplasias in the female, properly so-called; the allantois wholly transformed into a bladder, which opens directly, without the intermediate urethra, into the uro-genital sinus—that is, into the vestibule.
any part of the generative tract, considered first, in general, as abnormalities of the external and internal zones; and, secondly, as abnormalities in individual organs, dividing them for consideration into:

1. General anomalies of the two zones: true and apparent hermaphrodisim.
2. Anomalies of the separate organs:
   a. The external zone: the vulva, labia, nymphæ, clitoris, and the vagina; hypospadias and epispadias.
   b. The internal zone: the uterus, Fallopian tubes, and ovaries.

I. General—Total Absence of Either or of Both Zones.

There is on record no authentic case of entire absence of both external and internal generative organs in the same person. Occasionally there have been reported cases of acephalic fetuses, prematurely born, in which no trace of generative organs could be discovered, but these are extremely rare; more than that, no authentic cases have been proven, although many have been described, in which the external genitals have been entirely lacking; in every case properly examined rudimentary processes have been found.

Foville reported a case in which there was absence of the nymphæ, labia, and clitoris, with a fusion of the vestibule; a minute opening only was present, the outlet of the urogenital canals, through which the urine and menstrual fluid passed. In this case Klebs claimed there was fusion of the raphé. Meckel has described some old cases of entire absence of the genitals, but in these cases there was a depression or an elevation where the vulva should have been, and the details of the examinations were so meagre that they cannot be called authentic cases. The complete absence of the internal organs of generation is an extremely rare anomaly, if it exists. Kussmaul describes a female in which the most careful examination showed no signs of uterus, ovaries, or tubes, and where the vagina existed as a minute opening. Emmet records a case where a woman, so called, had been married for two years, but had never menstruated. An examination showed that sexual intercourse had been carried on through the urethra and into the bladder. In this case he was unable to discover any signs of vagina or uterus.

Other writers have described similar cases, but in few of them
has an autopsy been obtained, and then, in each case examined, rudimentary organs have been discovered.

True Hermaphrodism, in which one or more of the generative organs of the male and female are present in the same individual.

Dohrn denies the existence of true hermaphrodism in the human race, however common it may be in the vegetable and animal kingdoms, while Skene mentions Hildebrant and Bannon as having authentic cases which they reported. Klebs classified hermaphrodism into—

1. Bilateral, where the ovaries and testicles exist simultaneously on both sides;
2. Unilateral, where both ovary and testicle are present on one side at least;
3. Lateral, where the ovary and testicle are present on different sides.

Ahlfeld claims that there has never been on record a proven case of unilateral hermaphrodism, and that he has his doubts about the existence of bilateral hermaphrodism. Zweifel agrees with him in this. Ahlfeld mentions the cases reported by Heppner and Schnell of bilateral hermaphrodism, but there was so much difference of opinion about them that certainly nothing definite was proven. Zweifel quotes the following men as having recorded cases of lateral hermaphrodism: Sue, Barkow, Berthold, Bannon, Meyer, Gruber, and Klotz. Courty divides true hermaphrodism into lateral, transverse, and vertical or double, and says: "Two cases are now recorded—one by Rokitansky and another by Heppner—which prove to a certainty that the simultaneous presence of organs, characteristic of both sexes, may be found in the same individual, not only the one on one side, the other on the other, but simultaneously on the same side." The autopsy in Rokitansky's case in 1869 showed two ovaries with their tubes, a rudimentary uterus, and one testicle, with vas deferens containing spermatozoa. This individual menstruated regularly, and had an imperforated penis and a bifid scrotum. The case of Heppner, the second one he reported, was the autopsy upon a six weeks' infant, in which he found a complete internal generative apparatus, a penis, hypospadias, and two supernumerary glands, which he pronounced to be testicles. Slavjansky declared that these two supernumerary glands were ovaries, and not testicles.

Zweifel says of congenital hermaphrodism: "In not a single
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case as yet, however, have spermatozoa been found in hermaphroditic, the ejaculations consisting simply of such a fluid as even

Fig. 35.

Pseudo-hermaphroditism proper. External genitals of Julia B.—(man). Feminine appearance of the parts with the penis raised and the thighs separated: b, frenum; m, meatus; ov, vulvar orifice.

females secrete on irritation of their sexual organs.” Still, it is certainly a fact that the tendency in the majority of cases is toward the male type, and that nearly all, if not all, authentic cases have been of lateral hermaphroditism. In apparent or pseudo-hermaphroditism the female may simulate the male type by an abnormal development of the clitoris and a hernial descent of the ovary into the labia, as described by Auger; or, in cases of hypospadias, the male may resemble the female, the fissure of the corpora cavernosa being taken for a vagina, and the penis, which in these cases is nearly always atrophied, being mistaken for an hypertrophied clitoris. In some of the cases described, the non-descent of the testicles into the scrotum made the diagnosis more difficult. Vice versa, Junie, Coste, Engel, and Hugnier describe cases of hypospadias in the female, with hypertrophy of the clitoris, that were regarded and reared
as males. Leopold recorded a case in which a male pseudo-hermaphrodite was married as a female. There existed, in place of the vagina, a cul-de-sac. Oldham cites two cases where herniated ovaries in persons who had never menstruated gave rise to a mistake of sex. Ricco and Steglehuer reported cases of the same sort.

II. ANOMALIES OF THE SEPARATE ORGANS—THE EXTERNAL ZONE; VULVA, LABIA, ETC.

Louis and Petit mention cases of acephalic monsters in which there was complete absence of the vulva. Two cases were described by Riolan in which the left labium majus was lacking. Kussmaul describes Rossi's case, where the vagina existed as a most minute opening, and Foville's case, referred to before, showed an absence of labia, nymphæ, and clitoris. Coste and Seggel have on record cases where the labia were undeveloped, being represented by little ridges of integument.
Meckel, Granville, and Mayer have cited instances where the labia majora have been rudimentary or lacking. There are, of course, many cases on record of a lack of development of the external genitals as a whole, and where the parts, even in adult life, resemble those of an infant. Cases of hyper-enlargement or multiplication of the labia are not so rare. Meissner, Morgagni, Winckel, and Neubauer mention cases where there have been three and fourfold labia and nymphæ. Zweifel quotes Halle as recording a case in which the nymphæ covered the anus. Among certain tribes (the Hottentots, for example) the labia are of enormous size and hang down for six or eight inches (the Hottentot apron).

Arnaud and Morpain describe cases of absence of the clitoris, and Mannosi refers to a case in which an autopsy showed no sign of even a rudimentary clitoris. Zweifel mentions Meissner as quoting unquestionable cases of congenital hypertrophy of the clitoris, reported by Tulpius, DeGraaf, Zachias Avicenna, Plater, Rhodius, and Panarali. Frick, Armand, and Coste report cases of hypertrophy where the clitoris was as large as an erect penis. Ahlfeld describes several cases of this sort in full. The clitoris, like all the other generative organs, may remain in an undeveloped state, and yet, according to some writers, may not be, properly speaking, an anomaly.

Congenital hypospadias and epispadias are not uncommon in the female. In epispadias the clitoris is split at its upper or lower portion, as the case may be. Roser, Schroder, Gosselin, and Testelin have reported cases. Roser's and Schroder's were operated upon and cured by Moricke and Frommel. In hypospadias the posterior wall of the urethra is lacking, the canal opening upward into the vagina. There is seldom a fissure of the clitoris in cases of hypospadias.

The Hymen.—Roze, in his interesting thesis, goes fully into the question of the abnormalities of the hymen, and Courty, in his work, discusses the question in full. Illustrations are given of the different anomalies.

Zweifel writes that "very likely, atresia of the hymen is not an anomaly of development," and quotes Briesky as expressing the opinion, that it is simply the secondary obliteration of a previously formed canal, through defective hornification of the superficial epithelium. Briesky in his chapter on congenital malforma-
tion says: "Hymenial atresia, however, is excessively rare as an acquired condition," and he states that he has but once met with a true atresia hymenalis in a new-born girl. The genitals were otherwise normal in this child. He gives an interesting list of eighteen cases of hymenalis and vaginal atresia operated upon and cured by him. Zweifel himself had a case of "atresia hymenalis" where the entire vagina was affected by this epithelial adhesion; so, too, the cases reported of double hymens are simply adhesions of the epithelial cells.

The Vagina.—Atresia or absence of the vagina may be partial or total, and, according to Courty, may coexist with absence of the uterus or with a normal uterus. Cook, Yagishita, Mattersdorf, and Barsony have lately recorded cases of congenital vaginal atresia. Atresia of the vagina, to quote Briesky, is probably due to a secondary adhesion, as is atresia of the hymen, rather than to an anomaly of insufficient formation. Bokal and Zweifel seem to agree with him in this theory. Briesky goes on to say that the arrest of development may be of two kinds—cloaca, due to defective division between the rectum and bladder, and the existence of intravaginal septa. The cloaca may be complete or incomplete; the latter may be uro-genital or recto-genital. "The atresia of the upper and middle portion of the vagina is due to the loss of the existing lumen of the divided or united vaginal portions of Müller's ducts," but when the lower vaginal part is wanting, there may be a total absence of the lower part of the Müllerian ducts. Courty describes complete uro-recto-vaginal cloaca in a new-born child, and a recto-vaginal cloaca in a girl of sixteen, who had an imperforate hymen and menstruated through the anus. He cites several other cases of cloaca more or less severe. There may be a transverse division of the vagina, the so-called double hymen, or a longitudinal division, either from right to left—a rare anomaly—or from before backward, the so-called double vagina. These divisions may be complete or incomplete. Puech states that more than one hundred cases of this anomaly have coexisted with anomalies of the uterus, and less than fifteen have been reported with a normal uterus. Great differences exist as to the length and breadth and shortness of normal vaginae; anomalies of excessive length, etc., have been described by Toison, Scanzoni, Courty, Zweifel, and Puech.
Internal Organs (the uterus, Fallopian tubes, and ovaries).—The Uterus.—The division of uterine anomalies is as follows:

I. Defectus uteri. Total absence of the uterus.
II. Rudimentarius uteri. Rudimentary uterus.
III. Uterus unicornis. The one-horned uterus.
IV. Uterus bicornis. The two-horned uterus.
V. Uterus septus. Two-chambered uterus.
VI. Uterus duplex or didelphys. The double uterus.
VII. Defectus et rudimentarius cervix uteri. Defective and rudimentary cervix of the uterus.

Abnormalities of position.

Borner gives as the probable ultimate causes of the faults of development in the uterus the following:

1. Interference with the approximation of union of the two lateral organs which go to form the uterus.
2. Interference with the disappearance of the vaginal septum formed by the union of the median walls, which gives the double-cavity uterus.
3. Nutritive disturbances in the original genital structure.
4. The fact that the obstacle to development may occur so early in fetal life that the foundations of a part of the uterine structure are not laid; in this way a segment on one or both sides may be missing. Hart and Barbour give as the two causes arrested development and arrested growth, which together operate to produce malformations.

I. Defectus Uteri.—Kussmaul and Borner claim that the uterus is rarely if ever entirely wanting, and that an autopsy on any case will reveal some vestige of a rudimentary or atrophied organ. Courty quotes a case in which there was claimed a total absence of the internal organs of generation. Borner, Quain, and Stegلهuer report cases in which, on the living subjects, they could find no trace of uterus, ovaries, or tubes. In monstrosities in which no uterus was found, no traces of the Müllерian ducts were discovered.

II. Uterus Rudimentarius.—Veit, Langenbeck, and Nega have described cases where the uterus seemed little more than a thickening on the posterior vesical wall. Cases have been reported varying from this highest grade of deformity to the approach of the normal. The ovaries in these cases are generally present, and are often normal; there is no trace in the more pronounced cases of any periodic ovulation. Borner, Tauffer, Langenbeck, and Peaslee report cases
where relief was sought for pains and backache occurring regularly each month, but without ever being accompanied by menstruation.

In a case of this sort Leopold operated and removed a rudimentary left uterine cornu and ovary with a perfect recovery.

III. *Uterus Unicornis.*—An anomaly in which only one horn of the uterus has been developed, the Müllarian duct on the opposite side being atrophied, absent, or undeveloped. In this case the uterus is elongated and lies, obliquely bent, to one or the other side. Pregnancy in these cases occurs naturally, if the vagina be normal, and the shape of the uterus causes the fetus to lie vertically. In a case of Moldenhauer’s, on delivery, rupture of the uterine walls occurred. Hegar, Frankenhausen, Borniski, and Borner describe cases where one cornu was atrophied.

Koeberle performed Cæsarean section and removed piecemeal a fetus from a right uterine horn.

Salin, Litzmann, and Sanger performed abdominal sections for
the removal of diagnosed dead fetuses, and found that in each case conception in a uterus unicornis had occurred.

**Fig. 40.**

**Uterus Unicornis with rudimentary cornu:** LH, Lo, LT, and L Lr, horn, ovary, tube, and round ligament of the left side; RH, Ro, RT, and R Lr, those of the right side.

IV. *Uterus bicornis* is the result of a non-union of that part of the Müllerian ducts which goes to form the body of the uterus, leaving a division or fissure, more or less pronounced, from before backward over the fundus, separating the cornu, which projects at a more or less obtuse angle, each cornu having its distinct cavity. The uterus in these cases is often twisted on its long axis, and may contain a partition-wall. Cases have been recorded in which the uterus and the cervix have been divided into two separate compartments. The two horns are seldom equally developed, but the ovaries and tubes are generally normal; the vagina, however, often has the same duplexity. There may be atresia of one of the horns. In cases of extreme separation of the two halves, menstruation does not always occur simultaneously from the two cornu, and in some cases a pregnancy in one half does not interfere with menstruation from the other. Henderson made interesting notes on a case of this kind, watching the woman for sixteen years and delivering her of six
children. In two or three of these pregnancies she menstruated during the whole term.

Gouterman reports a case in which pregnancy occurred in each horn separately and at different times.

V. *Uterus septus* is a uterus normal in shape and generally in size, but internally divided into two cavities by a partition. This partition may be complete, extending from the external os to the fun-
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dus, or may be incomplete and only extend part of the way. In this anomaly the ducts of Müller have coalesced, but the partition-wall has not been absorbed. Blackwood recorded a case in which menstruation occurred alternately from either side. This abnormality interferes very little with pregnancy, but if the placenta is attached to the thin partition-wall, profuse hemorrhage may occur. Ruge recently split the partition-wall in a woman who had miscarried twice, and in the third pregnancy she was delivered at term.

VI. Uterus duplex or Didelphys is the development of two complete and independent uteri, with no partition-wall and no adhesions. Mayrhofer claims that this anomaly can only occur with changes that would render life impossible, and so thinks that cases reported as duplex are only cases of septus.

In Olliver's interesting case the autopsy showed two distinct uteri, separated from each other by folds of the intestines; and Olliver quotes Bonnet as having had the same sort of a case. Heitzmann's case was similar to this, with the additional fact that not only the bodies of the two uteri, but also the two cervices, were widely separated. In all these cases there was but one set of appendages to each uterus, and but one broad and lateral ligament.

Winckel and Cassau have reported similar cases, and Schroder one in which the rectum was between the two uteri. Menstruation has been in these cases normal. Satschowa reports a case where both cavities were gravid at the same time.
VII. Abnormalities of the cervix uteri, are common both in the size and shape of the canal and the external os, and in the size and shape of the cervix itself. These are well described by Courty in his work on the uterus, ovaries, and tubes. Winckel and Heitzmann have recorded cases of a double os uteri, or a normal uterus and cervix with the external os divided into two parts by an unabsorbed partition. Borner describes a case of his own of complication of the cervical cavity, which appeared at first sight like a cervix within a cervix, and quotes a case of Breisky’s at Berne which was of the same kind. Borner was the first to describe this anomaly, and considers it extremely rare.

VIII. Abnormality of position is caused probably by the insufficient development of one of the Müllerian ducts, although united to its opposite duct; again, there may be a difference in the position of the two Müller’s ducts, one being lower than the other, so that the fundus when developed is bent to the right or left as the case may be, or even twisted upon itself.

Kussmaul found this malposition in an autopsy upon a child, and in his case, one of the lateral ligaments was abnormally short. Fetal inflammations may play their part in these abnormalities. Sterility generally is present in these cases.

The Abnormalities of the Fallopian Tubes.—The entire absence of the Fallopian tubes rarely occurs, Courty says, even when the uterus is entirely absent. In cases of uterus unicornis, both the tube and ovary are lacking on the undeveloped side. Winckel, in post-mortem examination of 500 female bodies, found the tubes to be of unequal length in 25; in 3 cases the tubes were from 4½ to 5 inches long; and in 2 cases he found accessory tubal ostia. Klob and Rokitansky have called attention to the differences in form of the ends of the tubes, and described supplementary openings that sometimes occur at or near the ends. Hennig described three cases of accessory tubes, and Bandl reported a case in which the tube was normally developed, but imperforated. Congenital abnormalities of position and development of the uterus naturally give rise to abnormalities of position of the tubes, and congenital hernias of the ovaries carry the tubes with them as a rule. Olshausen says: “In some, the Fallopian tube is defective, and its internal extremity is alone developed; its abdominal extremity is destitute of fimbria and obliterated.” Keppler describes a supernumerary tube with a corresponding third ovary, that occurred in one of his cases.
Ovarian Anomalies.—Congenital absence of both ovaries, like absence of both tubes, probably occurs only in non-viable monstrosities, according to Olshausen, and reported cases in individuals cannot be considered authentic, since torsion and constriction may cause such marked atrophy as to leave little, if any, vestiges of the once-present ovary. Rokitansky demonstrated this condition in several of his cases. Absence of one ovary occurs only in cases of uterus unicornis. Grohe first reported a case of supernumerary ovary, and mentions a second case described by Klebs where the constriction of a band cut the ovary into two halves, each containing Graafian follicles in a rudimentary state.

Sinery’s autopsy on a new-born babe showed six appendages to one of the ovaries: one of these appendages showed normal ovarian structure, while the rest were cystic.

Keppler, as mentioned before, found a third ovary and tube in one of his cases. Kochs, Lummiczer, and Winckel describe similar cases. Beigel found appendages to normal ovaries containing ovarian tissue 8 times in 350 post-mortems, and Winckel 18 times in 500 autopsies. Waldeyer found 6 in one ovary. These extra ovaries are generally bilateral; their peculiar feature is their imperfect development. Klebs declares that ovaries, in which germinal epithelium projects into the stroma, with separation of these tubes from the surface epithelium, without the development of follicles and ova, are similar in many ways and in appearance to testicles.