CONGENITAL ABSENCE OF THE UTERUS.

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I.

In November, 1895, I reported[3] a case of congenital absence of the uterus, the first I had encountered in about 4,000 women personally examined by me. Since then I have seen in hospital clinical experience three additional cases, making four cases in somewhat over 10,000 women that I have had occasion to examine. This gives for me a frequency of one case in about 2,500 women, a percentage which I believe to be unduly large, since we as gynecologists see only those women who believe that they are not sound in their genital organs and who come for a digital exploration. Moreover, one man will see incidentally more of a certain rare condition than another of equal or larger experience. Thus, a gynecologist of large experience who has examined many thousand women informs me that he has never seen a case of congenital absence of the uterus. The true percentage, I believe is nearer one case in 5,000 than one in 2,500 women. Burrage[2], in 1897, collected references to 350 cases of absence of the uterus reported by 239 authors from earliest times up to that date, and, roughly estimated, about 300 of these cases were noted in the last century. This would seem to indicate even a smaller percentage than I have suggested.

Case 1.—The history of the case I reported is as follows: Clara F., a Russian girl of rather pleasing cast of features, and decidedly more intelligent than most of the women exhibiting the same anatomic condition, presented herself at the clinic of Prof. Harris A. Slocum, in the Polyclinic Hospital, on the evening of April 9, 1895. She stated that she was 18 years of age. She was somewhat undersized, but fairly well nourished. Her general complexion was dark, and her appearance mature. In no way did her physique suggest the existence of any physical defect. Her voice was full and mature, her manner timid. She further stated that she had never seen her menses, that she suffered considerably from headaches, mainly frontal in situation, that her habit was markedly constipated, that she was subject to severe cardiac palpitation, and that she often complained of vague, low-down abdominal pains. As far as could be ascertained she did not at any time suffer from menstrual molimina. Stethoscopic examination of the heart revealed a loud murmur associated with a condition of mitral stenosis. This cardiac condition was very materially improved under the exhibition of a mixture

consisting of the fluid extract of cactus grandiflorus, strychnin, compound syrup of the hypophosphites, and compound tincture of gentian. The action of the heart under this formula became more regular, and the distressing symptoms largely disappeared. On investigating the cause of the amenorrhea, an interesting state of affairs was discovered. It was found impossible to introduce the finger into the vaginal tract, although the vulvar orifice appeared to be normal in every respect. The pubic hairs were fairly well developed, but there was a complete absence of hairs in the axillæ. The mammæ were no larger than those of a man. Rectal exploration of the pelvis was then resorted to, but no trace of a uterine body could be detected, the finger of the hand placed above the pubic symphysis coming in contact with the rectal finger in all directions. This examination did not elicit any tenderness, and the abdominal walls were remarkably lax for a virgin, thereby still further facilitating the exploration. The uterine sound introduced into the vagina reached the upper terminus of that organ one and a half inches above the vulvar orifice. The woman was apprised of her condition, but she remained under observation for a few weeks only.

The additional cases are now reported for the first time.

Case 2.—Yedda W., a Russian girl, 20 years of age, presented herself at the clinic in the Polyclinic Hospital in February, 1897, to ascertain why she had never seen her monthly sickness. In sharp contradistinction to the foregoing case this young woman was coarse and repulsive looking. She had the flattened nose and heavy features of certain of the Russian peasant class, and her voice was harsh and strident. She was short and clumsily built, and gave the appearance of insufficient general development. She stated that her menses had never appeared and that she had at no time experienced any of the menstrual molimina. An examination of the chest showed a lack of development of the mammary glands, which were no larger than those of the ordinary male individual. There were no axillary hairs; the pelvic contour was contracted and closely approximated that of the male. There were a few straggling pubic hairs. The labia majora were small and flaccid and the labia minora very much under size. An attempt at digital exploration failed. There was a small canal about three-quarters of an inch in depth, barely admitting the index finger and ending in a cul-de-sac without a cervical projection. Subsequent bimanual exploration performed during etherization and with the left index finger introduced into the rectum showed an apparent total absence of the usual pelvic viscera. The patient stated that she experienced none of the normal attraction

to the opposite sex. As is usually the case, the girl disappeared shortly after she learned of her true physical condition.

Case 3.—Millie Dariana, a tall and attractive-looking Italian girl, aged 18 years, came to the clinic at the Pennsylvania Hospital on the 14th of September, 1901. She stated that she had been married six months, but that the marital relation had never been possible and she had come to learn why this was. Inquiry elicited the fact that she had never menstruated, but that every month she suffered from moderately severe pain in the lower abdominal region, but without cramps. Her sister, older than herself, had menstruated first when seventeen years of age. To all appearances this girl was normally developed. Her breasts were full but not large. There was a normal growth of axillary and pubic hair. The labia majora were of average size, and the nymphæ small but well marked. The rudimentary vagina had a depth of but one-half the length of the first phalanx of the index finger and ended, as usually, in a small cul-de-sac. Bimanual examination *per rectum* showed an apparent absence of the internal organs of generation, the tips of the fingers of the two hands coming readily in contact at every point through the abdominal and rectal walls. Upon being informed of her condition, the patient left and did not return.

Case 4.—Lucy Benfouri, an Italian girl, 19 years of age, presented herself in the dispensary service of the Pennsylvania Hospital on the 28th of October, 1902. She was single and had never menstruated, but every month for the past year had experienced some pain in the left inguinal region, the pain persisting for two days. She had frequently suffered with headache during the past seven months, accompanied with vertigo, malaise and anorexia. The girl was anemic and under size, not more than four feet ten inches in height, and generally poorly developed. She was a dyspeptic, a sufferer from marked constipation, and complained of palpitation on exertion. There was a slight growth of pubic and axillary hair; the labia majora were but poorly developed, and there was no trace of the labia minora. The urethral orifice was situated at the upper angle of a triangular expanse of what was believed to be an inperforate membrane occluding the vaginal orifice. Rectal examination, however, showed that there was no trace of vagina, uterus or other pelvic organ. The administration of Blaud's pills and arsenic had largely corrected the anemia. The patient denied the existence of the genital sense, and promptly disappeared after learning her true condition.

Since Burrage's paper was published there have been, as far as I

have been able to ascertain, but two other cases of congenital absence of the uterus recorded in the world's literature, as follows:

McCann[6]: A case of entire absence of all generative organs, vagina included; the pubic hair was normally developed.

Leech[5]: Complete absence of the vagina and uterus.

While preparing this paper the following letter containing the records of another case as yet unpublished, was received in the editorial office of the *Philadelphia Medical Journal:*

To the Editor of the *Philadelphia Medical Journal:*

Recently a Karen woman, about 20 years of age, came to me stating that her menses had never appeared, and that, although married for three years, she had never conceived. I made an examination and found externally the appearance of the parts were normal. On attempting to introduce my finger into the vagina, I found it entirely obstructed by a corrugated slightly elastic mucous membrane. I examined closely with a probe for any perforation, but none was apparent. After the bowel had been emptied by enema, I made an examination per rectum, and failed to find the slightest trace of the uterus or ovaries or any body simulating them. I therefore came to the conclusion that the woman was devoid of vagina, uterus and ovaries. She complained of no discomfort, and the reason for the desire for the examination was due to the husband's wishing to know the exact condition of things.

Very truly yours,

E. S. CORSON, M.D., of Toungoo, Burma.

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II.

The etiology of conditions such as this must necessarily be more or less obscure. It is generally admitted as beyond controversy that both the uterus and Fallopian tubes are derived from the embryonic Müllerian ducts. Williams,[7] in his recent admirable work, states that "according to His, the first signs of their development can be noted in embryos having a body-length of from 7 to 7.5 millimeters, when a thickening may be noticed in the celomic epithelium on the outer margin of each Wolffian body. These gradually become converted into two epithelial ducts, which converge and eventually meet together in the middle line, terminating in the urogenital sinus. The Müllerian ducts reach the urogenital sinus in embryos having a body-length of 2.5 to 3.5 centimeters. Their upper ends form the Fallopian tubes, while their lower portions fuse together to form the uterus and vagina. The fusion of the Müllerian ducts is usually completed at about the

third month, though the point at which the process is to occur is indicated at a much earlier period by the position of the round ligament." It is plainly to be seen, then, that owing to their distinct origin ovaries may be present in the absence of the uterus and vagina, and patients suffering from this curious defect may present all the menstrual molimina, including ovarian dysmenorrhea, backache and general malaise, as well as possess a certain amount of the genital sense.

Now, as to what factors are at work in the early weeks of gestation to prevent the development of these ducts and their ultimate fusion, nothing definite is known. An interesting theory has been recently advanced by Ballantyne[1], who would claim that all congenital anomalies, such as absence or rudimentary development of organs, or double formations, all of which are arrestments of normal embryologic processes or disturbances of embryogenesis, are brought about by the action of traumatism, microbes, or toxins upon the embryo *in utero*. While this has not been absolutely demonstrated, it is a very plausible theory, and one which may be accepted until controverted or supplanted by a better. Be this as it may, it remains true that the variety of the congenital defect will depend upon the time in embryogenesis at which the disturbing factor becomes operative. Most commonly, judging from the comparative frequency of the varieties, this occurs late in embryonic life, after the ducts of Müller have attained their full maturity, but prior to the time at which they have fully coalesced to form the generative organs. The various forms of double uteri and vaginæ are thus evolved. If, however, the arrest of development occur prior to the formation of the uterus by fusion of these ducts, or prior to the development of the ducts of Müller themselves, either one or both of these structures fail to appear; in the former instance there results a uterus unicornis, or one formed by but a single Müllerian duct; in the latter case no trace of the uterus can be detected on manipulation, even when the patient is completely relaxed under the influence of an anesthetic. If, however, the pelvic cavities of such individuals could be examined carefully after death, it is not improbable that in almost all, if not in every case, some trace of the missing structures could be detected, microscopically if not macroscopically, in the form of fragments of rudimentary muscular tissue. In the 360 cases collected by Burrage, there were but 35 autopsies, 24 of which were on the bodies of adults, 2 on girls, 10 and 12 years old, respectively; the rest being on monstrosities and fetuses with absence of other organs, making prolonged life impossible. In all of the autopsies on the bodies of adults and girls there were noted in every case rudimentary tissues representing the

uterus, generally occurring as one or two little knobs of tissue the size of hazelnuts or as a thin band of muscular tissue lying between the rectum and bladder. The ovaries were found to be present in all but six; the tubes also were present in all but six cases, though often without a canal.

The grouping of these congenital defects as proposed by McCann is as good as any yet suggested, and is as follows: *1st.* Cases in which no uterus is present, the vagina being a cul-de-sac. (In this class are to be found the first three cases reported in this paper, the last case being even a more pronounced abnormality, or one resulting from an earlier beginning of the disturbance of embryogenesis). *2nd. Cases* in which, at the blind end of the vagina, a nodule is found occupying a central position; this nodule has no canal in its interior. *3rd. Cases* in which the nodule forms the lower portion of a small central body admitting the point of a probe. *4th.* Cases in which the probe can be passed for a distance of from ½ to ¾ of an inch into a body, cord-like or fusiform, occupying a position corresponding to the normal uterus. This group forms a transitional stage preliminary to the so-called *infantile uterus,* which is the normal condition in the female child at full term, and which may persist through adult life if the normal processes of post-natal growth are retarded. Jacobi,[4] who has made a close study of these abnormal conditions, concludes that there may be noted the following varieties of genital defects in women: *1st.* Absence of the uterus with rudimentary development of the external genital organs (atrophy of the middle segment of the genital apparatus with imperfect development of the external segment). *2nd.* Absence of the uterus with hernia of the ovaries. *3rd.* An ambiguous and rudimentary condition of the external organs; the internal organs being masculine. *4th.* Ambiguous external organs, habitus masculine, the nature of the deep gland being uncertain. *5th.* External organs masculine, the internal feminine, habitus feminine. *6th.* External organs feminine, internal organs masculine. *7th.* The middle segment of the generative organs bisexual, the external masculine. *8th.* Bisexualism of the three segments.

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