THE DUPLICATION

OF

MALE AND FEMALE

EXTERNAL GENITALIA:

With Records of Two Cases.

by

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THESIS for the Degree of M.D.

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INTRODUCTION.

The rare cases of malformation that have their origin in the embryo present interesting problems. Their discovery compels our thoughts to revolve on the all-absorbing study of the de-:velopment of the embryo, which has still many questions left unanswered. Although the solu-:tion of these problems of development lies in the hands of the skilled embryologist, no one can give more valuable aid than the general practitioner. In his obstetrical work he can-:not fail to be interested in foetal malforma-:tions, for it is into his hands that speci-:mens illustrating these conditions in the first instance pass and to him falls the oppor-:tunity of examining the placenta, membranes and umbilical cord. He must ever appreciate

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the possible nature of material which may come his way and realise that, by actively co-operating with the embryologist, he may by chance provide the solution to some of the mysteries of the early development of the human embryo.

The general practitioner has a great responsibility in making known the rare cases of congenital malformation. He is constantly in touch with the new generation and has therefore the unequalled opportunity of making the earliest observations on any case that shows a departure from the normal type. Further, he is in a position to pursue enquiries into the history of progenitors where the question of heredity arises and so furnish valuable data in the study of the mysterious phenomena of heredity which is closely allied

to/

to the study of congenital deformity. "It would be an advantage" says Cooke Hirst (1) "to begin the observation of the foetus, as well as its treatment, a hundred years before its pro-:creation, in a study of antecedent generations".

If the general practitioner can find the leisure to pursue the study of the perplexities involved in any case of malformation which may come his way, or, if unable to do so during the busy routine of his duties, he even passes on his material and records to those who can probe the deeper aspects of the problem in the light of more specialised knowledge, he has made his contribution to science.

In the general practitioner's life there is so much of interest and importance to be done and opportunities are many. Yet time is so short/

short that we are in danger of forgetting that our duties end, not with ourselves in making a living nor with our many patients in doing our best for them, but extend to an obligation to the profession of making some more permanent contribution, small though it be, to the sum of human knowledge.

An opportunity unique in the history of medicine has led me to describe and discuss two cases of Duplicated External Genitalia, a rare type of developmental abnormality. There is no record of any one medical man in his lifetime having encountered two similar cases. Not only was it my good fortune to attend at the births of the children but the parents in both instances have been patients of mine for many years. My personal knowledge of them

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and of their relatives has enabled me to fur-
:nish an authentic genealogical tree of out-
:standing interest, in itself an important
link in the solution of the explanation of
the malformations.

With Duplicated External Genitalia, as with a great number of cases of malformation, there are not infrequently associated multiple defects. A study of such cases has elicited valuable information regarding the factors concerned in their production and has often served to throw light on the intricate pro-
:blems of development.

A.

D U P L I C A T I O N

of

M A L E E X T E R N A L G E N I T A L I A.

HISTORICAL NOTES.

By far the greatest number of malforma-:tions can be traced to the early period of gestation and the rarest type of these is duplication of external genitalia. The out-:standing features of twenty-four cases on record of this deformity occurring in the male are here considered.

The first case of Double Penis was seen in 1609 by G. G. Wecker (2). The second was seen in Belgium and reported by T. Bartholin (3) in 1657. The third was seen by M. B. Valentini (4) in Germany in 1696. These three were the only cases recorded in the seventeenth century. Except for the fact mentioned by Bartholin regarding his case that the man was exhibited publicly because

of/

of the presence of two penises which lay side by side the recorders furnished no details of their cases.

During the eighteenth century only four cases can be traced. At the beginning of that century G. Ollsner (5) reported the in-:teresting case of a man who was first seen when he reached the age of twenty-two and was a soldier. As is the case with a number of cases of double penis, the man's condition was really bifid penis. The penis was divided in the neighbourhood of the glans and urine was passed by the meatus in the left glans. Because of the deformity the man was incapable of true sexual intercourse.

In 1740, Hahnius (6) reported a case of double penis but no details of the case were given/

given.

In the same year Becchetonni's (7) case was seen in Bologna. It was stated that the two penises lay side by side.

The only other case in the eighteenth century was that of Niemann (8) recorded in 1774. It was seen in Germany and the record of the case contains the following statement:- "Urine and semen were passed from both penises and the man's wife always gave birth to twins".

At the beginning of the nineteenth century Loder (9) mentioned without proper record, a case seen at Gottingen in 1802 and in 1813 F. Sixtus (10) published at Wurzburg several details of a case which came under his observation. The case was that of an old man, who had actually reached the mature age of ninety-three/

ninety-three. The penis was split into two in more than half its length. He states that in the uppermost part of the scrotum there was a fissure which communicated with the bladder.

In 1837 Jenisch (11) described what was up till that time the most interesting case that had been discovered. The two penises lay side by side and each had a patent urethra and its own scrotum but each scrotum had only one testicle. The anus was imper-:forate.

In 1846 Wm. Acton (12) in London examined and described the famous case of John Baptist Dos Santos, a native of Portugal. As the child presented peculiarities which the medical attendant thought unique, the parents determined/

determined to come to London to exhibit the child, who was described in a printed paper as "The three legged child with the first bipenis ever seen or heard of". Each penis had its scrotum, the outer half of each con-:taining a testicle. Urine was passed from both penises simultaneously. The anus was in normal situation. The interesting fact is also added that "Between and behind the legs there was a third extremity or rather two lower extremities united in their whole length". The foot of this extremity was double and was supplied with ten toes, the two great toes occupying the centre position. The author stated that this instance of two penises on one body did not result from a division of one organ, the distance apart of

the penises and urethrae making this little probable.

In the same year, 1846, Pigné (13) who was the curator of the Dupuytren Museum in Paris demonstrated a case of double penis in support of his theory that duplication of organs resulted from fused germs. In this case the author described a small penis below a normal penis. Dissection showed on the external surface of the bladder a groove which indicated a commencing division of the bladder into two and was really an incomplete septum. The rectum was single but there was duplica-:tion of stomach, heart, lungs and aorta, the two aortae fusing at the level of the fourth Dorsal Vertebra.

Buren and Keyes' case (14) recorded in

1873/

1873, was a man of forty-two who had two distinct male organs which were described as being of normal size. They were well-formed and lay in apposition, with a common skin covering, extending as far as the base of the glans. The meatus of the left glans was blind and urine escaped through the meatus in the right glans and also from a small ori-:fice in the perineum. "He was compelled to let down his trousers and sit when he made water". The pelvis was broader than normal.

Sangalli's (15) case was described in 1875. The patient died in 1894 and for the first time in the details of the records of a case of double penis the full account of the post-mortem examination is given. The man was aged thirty-five. The interesting points recorded/

recorded were that the pelvis was wide and defective in the middle line in front and the man had a staggering gait. A single deficient scrotum was present. The two penises lay widely apart and the two bladders which were present intercommunicated and opened to one urethra while the other urethra was blind. The position of the anus was abnormal. This is the first case of double penis recorded where the rare abnormality of double bladder was present.

In 1878 A. P. Smith (16) published in America a most interesting case. He was consulted by a patient who complained of severe pain on micturition. Examination revealed a double penis and each penis communicated with a bladder. His symptoms were discovered to

be/

be resulting from the presence of stone in one of the bladders while the other was in a healthy condition. The operation for the re-:moval of the stone was successfully performed.

In 1887 Taruffi (17) the Italian Terato-:logist, fully described two cases which had been referred to him. The first was a child of three and a half years, the third child of healthy parents. It possessed two penises, each with a patent urethra, and two well-:formed scrotal sacs. The pelvis was large and there were two anal orifices. This is the first recorded case of double penis accompanied by the very rare deformity of double anus.

Taruffi's second case (18) was that of a man who had attained the age of forty before the/

the malformation of his external genital organs came under notice. "The patient stated that both penises became erect at the same time and that he had not married in order to conceal his condition".

In 1894 Dr. Cole (19) in America re-:corded his case of a newly born baby which weighed 10 lbs and was supplied with two well-:developed male organs, half an inch apart. There was an imperforate anus.

In the same year Dr. Scot Skirving of Edinburgh (20) described the first case in this country. The scrotum was divided by a deep groove in the middle line into two dis-:tinct lobes. At the lower end of the groove there was a swelling, from which arose two short but perfectly recognisable penises projecting/

projecting upward and outward. Both of these, instead of having a long foreskin, had a short prepuce exposing the whole glans and laying bare the whole edge of the corona. Each penis had a distinct meatus urinarius but that on the left side appeared to end blindly. There was a deficiency in the integument of the perineum and in the centre of this there was a granulation mass, from which urine escaped. The anus was in normal position. No post-mortem examination was made.

In 1895 Dr. Lange (21) of Leipzig pub-:lished a case in which there were two well-:formed penises, each communicating with the respective half of a urinary bladder. The bladder was divided into two equal parts

by/

by a median perpendicular fold. There was no sign of division externally and there was only one urachus present. During life meconium had been passed from both penises and there-:fore a communication existed between the membranous part of the urethra and the rectum. The scrotum was divided into three parts by two raphés which passed from below the root of the penis. The lateral portions contained a fully formed testicle. There was no in-:dication of an anus or any duplication in the skeleton, especially in the spinal column or pelvis or in any of the internal organs.

In the same year, 1895, J. H. Morgan (22) gave a short description of an infant of eighteen months. On each side there was a large and well-formed mass of scrotal tissue containing/

containing well-formed testes. The masses of scrotal tissue were separated at the upper extremity by two well-formed penises. There was no urinary meatus in either but in the perineum there was a mass of tissue, through which urine escaped.

J. A. Pires de Lima (23) in 1915 described a case of bifid penis in a man of fifty-eight. The organ was short but had a double well-developed imperforate glans. The urethra which was absent in its greater extent finished in a rounded meatus in front of the scrotum. The prepuce was double and extremely short. The scrotum with its contained testicles was normal. The man was a farm labourer and was married. The malformation of the genitals did not affect the sexual functions. There were two

daughters/

daughters and three sons of his marriage. No similar anomaly was known either in his ancestors or relations.

The author gives an embryological interpretation of the condition. He regards the genital tubercle as a double organ re-:sulting from the fusion of two symmetrical halves. "The penis has the value of two formations which, developing one by the side of the other, join together so as to form one single organ. The junction of the two primitive halves did not take place completely in the present case. The two corpora carvernosa, joined at the prepubic angle, separated afterwards ending each in an in-:dependent glans."

In 1924 A. MacLennan (24) of Glasgow described/

described a case of double penis. There was complete hypospadias of both penises. MacLennan offers an explanation of the abnormality. He says that "it cannot be regarded as an instance of arrested develop-:ment but rather should be considered as an unbalanced growth. The nature of the con-:dition may be taken to be a connecting link between an incomplete twin and an arrested development: in other words an error in the distribution of the trophic force pertaining to the anlage of the region involved. The anlage, which should have formed the genital organs, had shown an attempt at the formation of a twin. This excessive activity was countered by the inability to complete the formation, thus leaving an example of arrested development/

development in the hypospadias".

In 1930 Charles Donald, London (25) chronicled a case of double penis. The penises were quite separate, the left being at a slightly lower level than the right. The left urethra was patent in its whole extent but, except for a small depression in the right glans, there was no evidence of a right urethra. The scrotum consisted of two separate halves each containing a testicle. The prostate was normal and there was no evidence of arrested development in the pelvis. There was no family history of any abnormality. "The limitation of the duplicity" he says "in that no extra hind limbs or double pelvis were found may be interpreted in either of two ways: the genital tubercle may have been the seat of

local/

local dichotomy - the teratogenic agent act-
:ing upon it while it was in a state of high
metabolism - or the teratogenic process has
originally been wider and there has been a
partial secondary fusion".

SUMMARY OF THE RECORDED CASES:

As the discovery of a malformation in the form of Duplication of External Genitalia would excite great interest, it is most likely that the cases, which have occurred, have found their way into literature. It is evident from that number that it is a rare abnormality. Yet, a survey of the cases reveals in most only a few details of the condition. In fact, in the earliest cases recorded it is surprising to find that mention only is made of its occur-:rence.

It is noticed that in some of the cases described the double penis claimed to be present was really a bifid penis, the penis being normal to a point where it split into two. We note the evidence of associated

arrested/

arrested development such as hypospadias, deficiency of the prepuce, glans and corona. In only thirteen cases were there two distinct and well-formed penises placed side by side. In three only does the much rarer form occur where the one penis was slightly above and anterior to the other (as in my case described later). So also, in only a few were there two scrotal sacs. In the majority there was no true scrotum.

In many no details of the condition of the anus or rectum are given. Atresia ani was an associated defect in some cases and in one case the unique condition of double anus was described. In two of the cases meconium was passed along with the urine showing a persistence of cloacal conditions.

Post-mortem/

Post-mortem findings, except in a few cases, are not recorded. The prostate gland and the bladder came in for scant notice al-:though it was mentioned in two oases that there was a median ridge in the bladder and in two other cases there was present the very rare anomaly of double bladder. The condi-:tion of the other viscera is not described except in Pigné's case, where there was re-:markable duplication in other organs. In the case of Dos Santos described by Acton there was a third lower limb between two normal lower limbs. This third limb was seen to be really two extremities joined to-:gether in their whole length.

The important question of family history finds a place in the description of no single case/

case recorded. There is therefore nothing to show that these cases of duplication of External Genitalia occur in families where malformations are common and there is no re-:cord of more than one occurring in the same family. In fact the absence of any informa-:tion on the important question of family history is regrettable and there is there-:fore lacking in the discussion of the cases a group of facts of the utmost importance in arriving at a solution.

The abnormality had no effect on the lives of the individuals as all the recorded cases, with one exception, survived birth. Eight of the cases described when first seen were of ages ranging from twenty-two to forty-five and one case was not discovered until/

until the subject had reached the advanced age of ninety-three. Practically all the cases showed sterility. As Ballantyne (26) says "The apparent anatomical exuberance is usually accompanied by physiological in-:competence".

DESCRIPTION OF AUTHOR'S CASE.

An infant with double penis and scrotum came under my notice in September, 1925. At that time I was called to attend Mrs. H-, who was in labour. She gave birth to a full-time male baby, her second child. It weighed 9 lbs. The mother was well during the whole of her pregnancy. The labour was normal. The placenta, membranes and umbilical cord showed no abnormality. At birth, the child was found to have a curious formation of the external genitals and on close examination this was found to be due to a double penis and a double scrotum, both well-formed, one slightly above and anterior to the other. (Figs. 1 and 2, photographs, p.p. 30 and 31). The child sub-:sequently passed urine from the upper penis only/

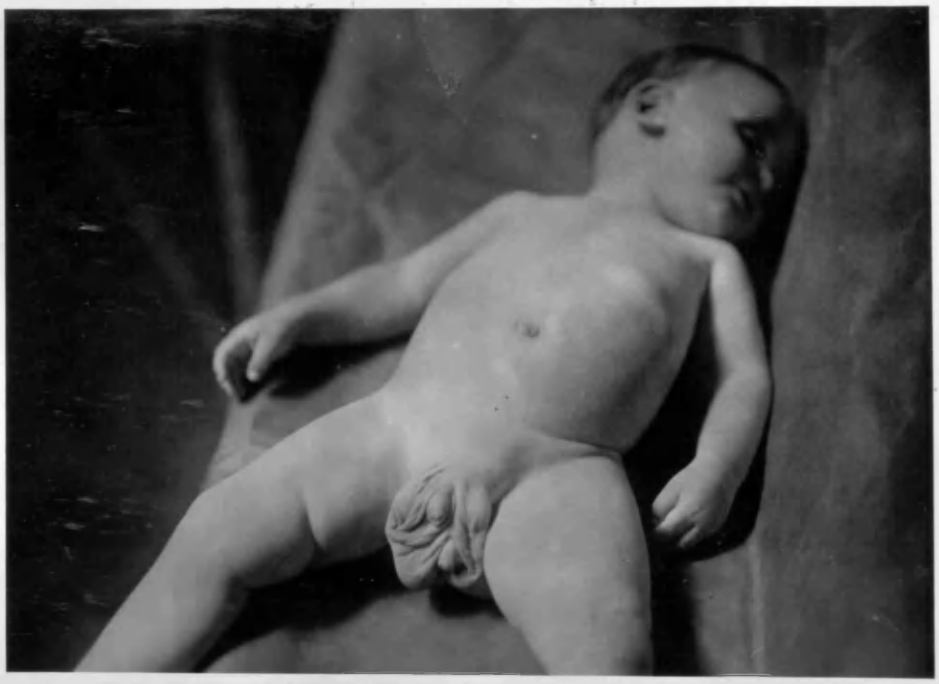


Fig. I. Photograph of Author's Male Case of Duplicated External Genitalia.

Fig. 2. Photograph of Author's Male Case of Duplicated External Genitalia - near view showing double well-formed penis and scrotum.

only. This seemed the functional one and was normal in form, size and attachments. There was a good corpus cavernosum but the glans was slightly deficient. The lower penis did not seem to function but appeared fairly normal except that the corpus cavernosum was small. Each penis had its scrotum. These lay side by side and to appearance were normal but palpation revealed only one testicle in each scrotum, one in the right half of the upper and one in the left half of the lower. The test-:icles were well developed, if anything, rather larger than normal. The raphé of each scrotum can be seen distinctly in the photographs. There was one natal cleft and the anus, with a well-developed sphineter ani, was in normal position. There was no deficiency in the integument/

integument of the perineum and although a careful search was made no other abnormality could be found in the child. The child was artificially fed and thrived with steady gain in weight. He cut his first tooth at seven months and had five teeth at nine months. At this age he was attempting to stand. Unfortunately he developed seborrhoeic eczema followed by acute nephritis. Ascites supervened and the child died during the night of 16th July, 1926, in a uraemic convulsion. He was then ten months old.

.....

Post-mortem Examination.

The child was big for his age and well-developed. The position of the umbilicus was normal. The double penis and double scrotum/

scrotum were of normal size, well-formed and separate in their whole extent. They were not situated on the same level but as al-:ready described occupied the very rare position seen in this deformity of being placed one slightly above and anterior to the other. Only two testicles were present. They looked larger than normal but the size was influenced by oedema resulting from the nephritis. The position of the testicles, one in the right half of the right scrotum and one in the left half of the left scrotum, as diagnosed on physical examination when the child was born, was here confirmed. The urethra on the right side was patent through-:out its whole extent but the left urethra was patent only in the penile portion.

Examination/

Examination of the abdomen revealed no abnormality. The peritoneal sac was normal as were the liver, spleen and kidneys, except for oedema and congestion seen generally in the condition of acute nephritis. There was a ureter on either side passing down to a single bladder of normal appearance. The interior of the bladder showed no abnormality. The outlet led down to the right urethra. There was no outlet or mark of an outlet for the left urethra. There was a single vas on each side and there were no undescended testicles. The bowel was normal and termin-:ated in an anus in the usual position. There was no malformation of the organs of the chest.

Although the abnormality of the penis

and/

and scrotum was an extraordinary one, post-
:mortem examination revealed the interesting
facts:-

1. The condition occurred in an individual otherwise normally developed.

2. The condition would have been quite amenable to surgical treatment, as both testicles were present and the upper functional penis was normal in form and attachments.

FAMILY HISTORY.

The parents of the malformed infant are both healthy and are members of large families. The father is now aged forty and is the tenth member of a family of ten. The mother is aged thirty-eight and is the twelfth member of a family of thirteen. At the time of the child's death I took blood from both parents for a Wassermann Test and in both cases the result was negative. There was no history of fright or traumatism to the mother during the pregnancy. Labour was normal and termin-:ated in the delivery of a well-developed and strong child. Previous to the infant's birth the mother had given birth to a male child who suffered from slight recurring attacks of seborrhoeic eczema but was otherwise healthy

and/

and has remained healthy. The mother has had a normal pregnancy and normal labour since, but the baby, a female, was a typical cretin. This child made a perfect recovery with thyroid treatment. (Figs. 3 and 4, photographs, p. 39).

There are two further points of interest in the family history:-

1. <u>The history of twinning</u> on both the maternal and paternal sides. The mother's sister had twin girls and the father's sister had twin boys. The twins occur in the same generation, are all alive and healthy, and it is noticeable that in both instances they are of the same sex.

2. The mother of the male child with

Fig. 3. Cretinism - Baby of 14 months - stunted growth, relatively short thick-set limbs and protruding tongue.

Fig. 4. Same patient after 8 months' Thyroid treatment.

Duplicated External Genitalia which has been described above is a full cousin of the mother of the female child with duplicated external genitalia, the second case to be described.

To avoid repetition discussion of this case is reserved until after a description of the Female case.

B.

DUPLICATION

of

FEMALE EXTERNAL GENITALIA.

HISTORICAL NOTES.

This is one of, if not, the rarest of malformations of its kind and, because of the associated duplication of other organs, one of the most interesting. Only seven authen-:tic cases can be traced after an exhaustive search into the literature. That number probably can be said to include all the cases that have ever occurred as it is inconceivable that a condition, so obvious and so interesting, should ever have passed unrecorded. The authors of the classical case in Great Britain describe their case as recently as 1913 and included in their description of it a brief account of what they considered the only other authentic one. But I have found seven cases which have been recorded and a short summary

of/

of their outstanding points is here given.

The first case of Double Vulva was re-:corded by Suppinger (27) in 1876. It was a case of a child who died when twenty-one months old. The description of it was brief. There were two separate vulvae. Post-mortem examination revealed a pelvis divided into two halves by a median peritoneal fold. In each half there was a bladder and rectum. "There were two separate coccyges".

In 1888 Bechlinger (28) reported from Para, Brazil, a case which closely resembled the male case of Dos Santos already described. The genitals were duplicated but she possessed "a third leg attached to a continuation of the coccyx and in addition to two well-developed mammae regularly situated, there were two rudimentary/

rudimentary ones close together above the pubes".

In 1906 E. Chill (29) recorded a case of an eighth month foetus, which survived only a few hours after birth. The whole of the abdominal wall was absent except for a small irregular flap of skin, to which was attached the umbilical cord. The external genitals were duplicated and separated by a central piece of skin. The double urogenital and anal orifices were extremely rudimentary.

In 1913 Gemmell and Paterson (30) of Liverpool fully described the classical case in this country. The woman was thirty-two years of age. There was no umbilicus and the pelvis had a wide appearance. There were two separate and laterally placed vulvae with/

with a single anus and a natal cleft behind the anus. The two vulvae were almost normal in size and form. The labia majora were well formed but the two inner labia majora were not so prominent as the outer. With each vulva were labia minora, clitoris, vestibule and the openings of the urethra and vagina. The openings of the vagina were three inches apart. The labia majora and clitoris were not so well formed on the right side as on the left. On each side there was a well-formed uterus and cervix. The urethrae were normal and there were two distinct bladders. "The pelvis is deficient in front in the middle line and a little to the right of the middle line there is a stellate cicatrix representing a pseudo-:umbilicus and the closure of the foetal

body-cavity/

body-cavity - at its junction with the placenta". The woman was pregnant on two occasions. On the first she was pregnant in the right uterus and gave birth to a male child at full time. On the second she was pregnant in the left uterus and gave birth to an eighth month female child which survived. In discussing the explanation of the abnormalities the authors point out that "The salient features of the case are; a single anus and rectum, duplication of vulva, urinary and genital tracts, separation of the pelvic bones and the absence of a true umbilicus". The authors hold that the abnormalities are accounted for by the absence of an allantois. Presuming that there is absence of the allantois, they maintain/

maintain, there is then no allantoic divert-
:iculum and no thick mesoblastic tissue to
carry the umbilical vessels. These are
carried to the chorion and placenta in the
mesoblastic tissue of the amnion. There is
therefore no umbilical cord. The rectum
and anus have developed normally but by the
absence of the allantois no normal bladder
is formed. Traction of the amnion has
favoured arrested development in the pelvic
girdle and has also favoured separate con-
:nection of the Müllerian ducts with the
External Cloaca and the formation of bi-
:lateral bladders and urethrae out of the
ventral part of the Internal Cloaca.

In 1924 MacLennan of Glasgow (31) re-
:ported a case of double vulva in an infant.

The/

The labia minora and clitoris were hyper-:trophied and the anus was imperforate. It was not known if two bladders were present. "The excessive trophic force, which resulted in the complete duplication of the vulva was countered by an imperforate condition of the anus".

In 1925 Blair Bell (32) described a case of diphallus in the female. Bifid penis is rare but this is the only case of the homologous condition in the female re-:corded in literature. The subject was a girl of fourteen. Examination showed "complete separation of the nymphae, clitorides and their preputia", with a median groove extending from the symphysis to the urethra. The vagina and uterus were normal/

normal and no abnormality could be proved in other organs. He considers the condition of diphallus to be due to the arrest of fusion of the bilateral elevations from which the phallus is developed. "Diphallus in man" he says "may be regarded as an atavism, rather than an actual malformation, for in certain of the lower creatures these duplex states are normal".

The seventh case of double vulva, was described by Dr. W. A. Hinckle (33) of Peoria, Illinois in 1928. The author kindly sent me the appended photograph of his case. (Fig. 5, p. 50).

The woman was aged sixty-two and had enjoyed normal health till the onset of in-:definite bowel and rectal symptoms. The abnormalities/

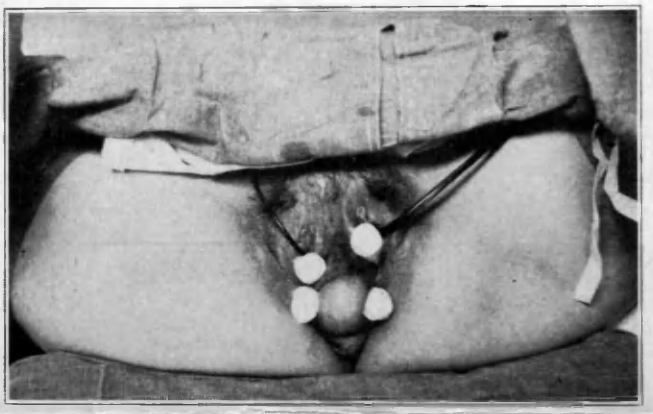


Fig. 5. Hinckle's Case (drawing sent to me by the author) - Double Anus, Vagina and Uterus. The white objects are pieces of gauze wrapped round the ends of rectal tubes and inserted into each rectum and vagina.

abnormalities present were unique. There were two definite vulvae but the labia majora on the inner aspect fused into a sort of perineal body. The organs of the left side were better developed than those on the right. There were two ani. Defaeca-:tion was usually through the left anus though, when the movements of the bowel were loose it took place through both and not always at the same time. Urination was from both urethrae and nearly always simultaneous. Before the menopause the patient menstruated from both vaginas though more freely from the left. Bi-:manual examination showed an apparently normal uterus on the left. The right vagina was small but a cervix was seen.

X-ray/

X-ray examination of the bowel was not very definite but showed two separate colons up to the caecum. The patient had been once pregnant and had a miscarriage of twins at the third month. No family history was elicited. Opportunity for post-mortem examination was not presented. No attempt is made to explain the abnormalities.

SUMMARY OF THE RECORDED CASES.

Double vulva is one of the rarest of teratological anomalies. It is much rarer than the corresponding condition in the male.

In only one case (the only one traceable in literature) was there described a partial duplication of external genitalia correspond-:ing to the condition of bifid penis. In all the other oases recorded the two vulvae were separate and usually quite well developed, although the labia majora on the inner aspect of the separate vulvae were less prominent than on the outer aspect. There was a tendency for the vulva to be better developed on one side than the other.

The condition becomes more interesting by reason of the remarkable duplication seen

in other organs. In four of the cases the uterus was double and in the same cases the very rare anomaly of double bladder was proved. Most of the cases of double bladder recorded in literature are of the nature of diverticula. The large bowel was duplicated in Hinckle's case.

Accompanying the condition was seen evidence of arrested development in parts of the external genitalia and anal orifice. The latter was entirely absent in two cases and extremely rudimentary in one. Double, well-:developed anus, an excessively rare condition, was present in Hinckle's case.

Suppinger's case was stated to have two separate coccyges.

It is interesting to note that the individuals/

individuals, with one exception, survived birth. Hinckle's case was first described when the woman reached the age of sixty-two and as already mentioned she had been once pregnant and aborted twins. Gemmell and Paterson's case was that of a woman of thirty-two who had given birth on two occasions to a living child.

In the records family history receives scant mention and post-mortem findings are conspicuously lacking.

DESCRIPTION OF THE AUTHOR'S CASE.

In June 1931, I attended Mrs. McP. in her first confinement. Her pregnancy was normal and she had sustained no injury throughout its course. The confinement was difficult owing to the presence in the baby of a huge lateral sacral meningocele, which extended low down into the left gluteal region. It was noticed at the birth that the child had complete duplication of external genitalia and double anus. The two vulvae were quite well developed although the outer labia majora were the more prominent. (Fig. 6, photograph and Fig. 7, drawing, p.p. 57 and 58). They were separated by an inch of unbroken skin. There were two vaginae. Two urethrae were also present and when the child was born urine

was/

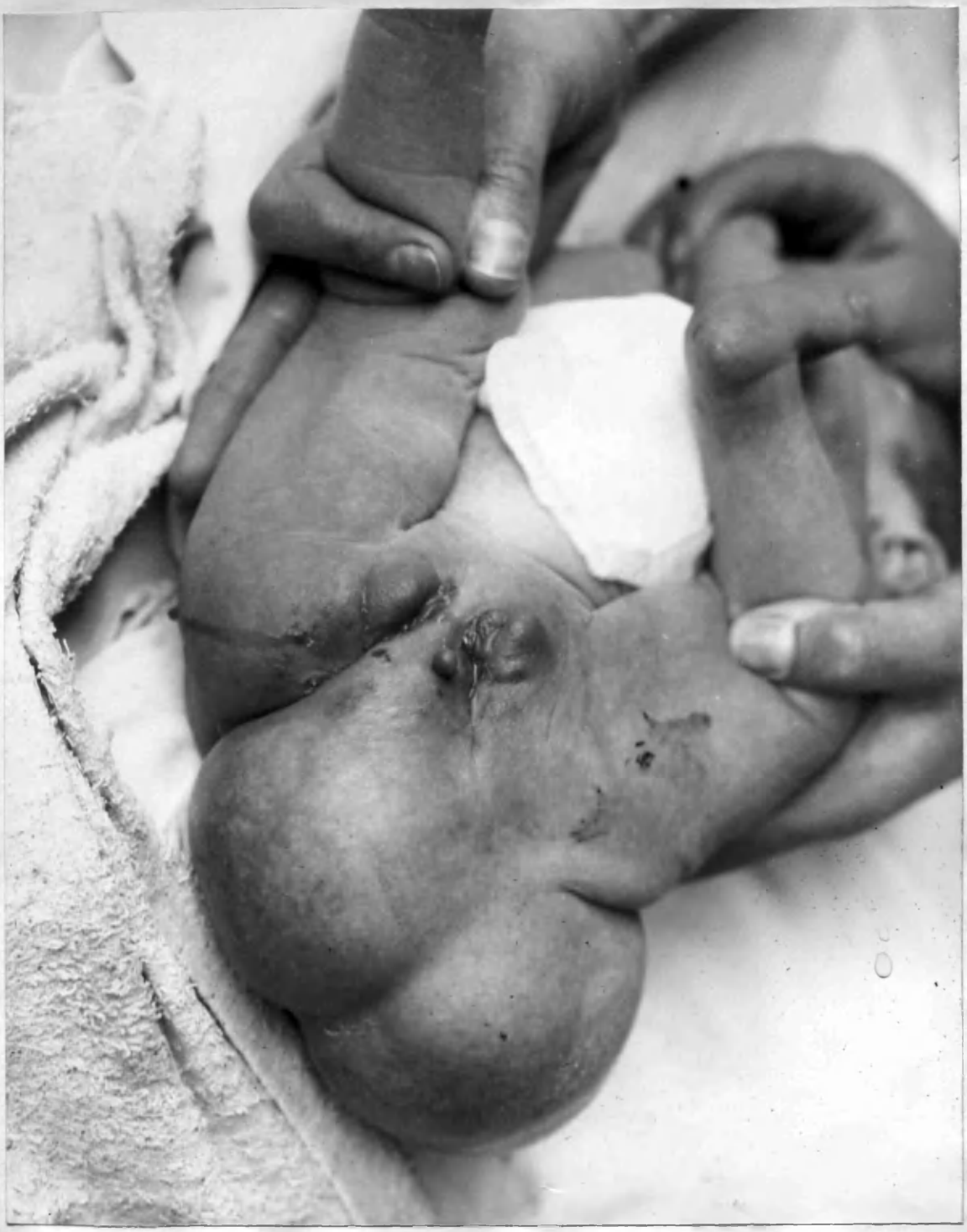


Fig. 6. Photograph of Author's Female Case of Duplicated External Genitalia - Double Vulva and more prominent left vestigial tail.

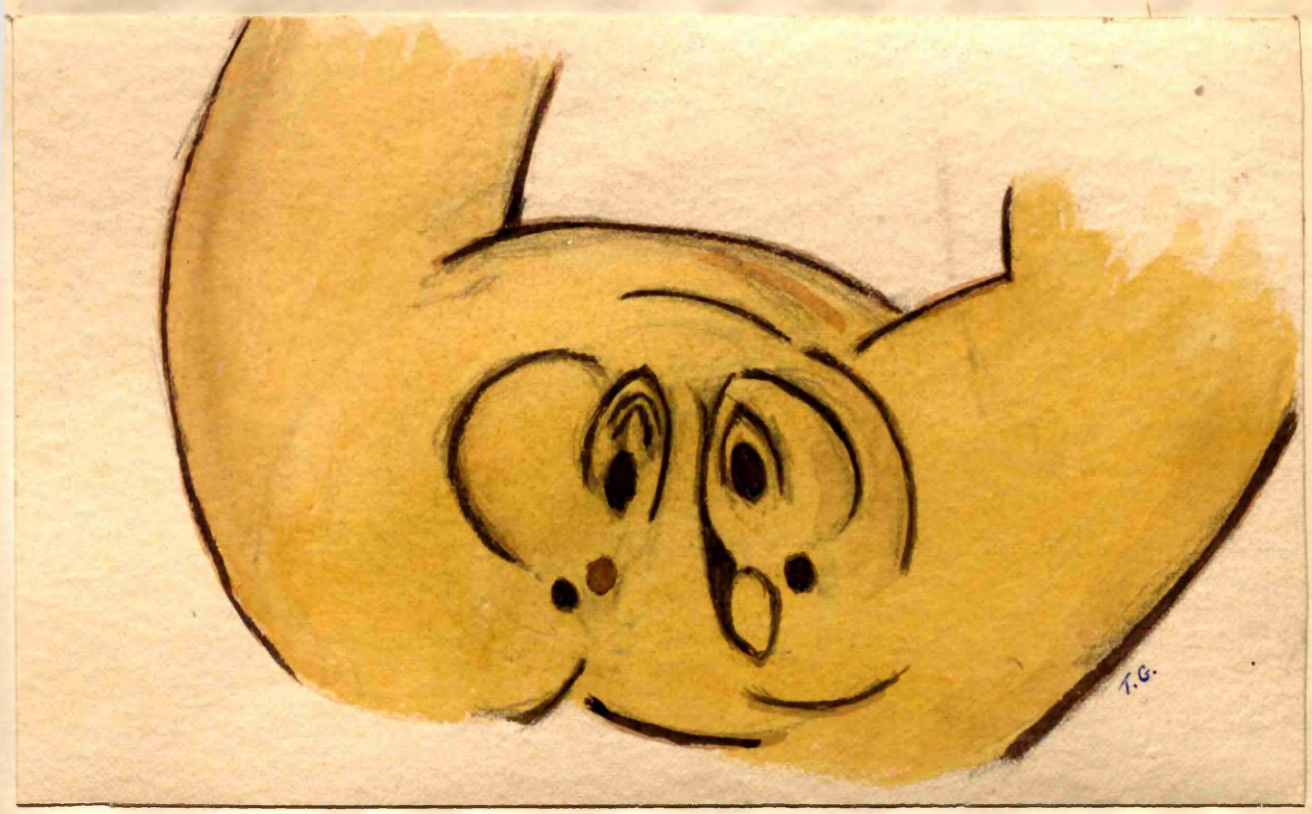


Fig. 7. Diagrammatic Drawing to show relative position of the parts more clearly.

was seen to escape from both simultaneously. The perineum was intact and there were two ani on the same level two inches apart with well-developed sphincters. Through both of these meconium was passed. Immediately to the inside of the left anus there was a small but distinct vestigial tail and on the right side there was a small elevation of skin in an exactly similar position which indicated the remains of a tail on the left side. There was a slight defect palpable over the symphysis pubis. The child weighed 8 lbs. and was slightly cyanosed. It was artificially fed and it was noticed that it often turned "blue" after feeding. It made poor progress but survived until the end of three months.

.....

Post-mortem Examination.

The post-mortem examination which was carried out for me in the Sick Children's Hospital, Edinburgh, revealed a series of remarkable and unique abnormalities. (Fig. 8, p. 61).

Double bladder was present. The bladders lay in apposition and were oval in shape. The walls were composed of a thick muscular and mucous layer. There was no connecting channel between the bladders. From an umbilicus, normal in size and situation, de-:scended a urachus, which on approaching the upper surface of the double bladder, broad-:ened out to form connection with the apices of the bladders. Microscopic examination revealed the urachus double. (Fig. 9, p. 62).

Both/

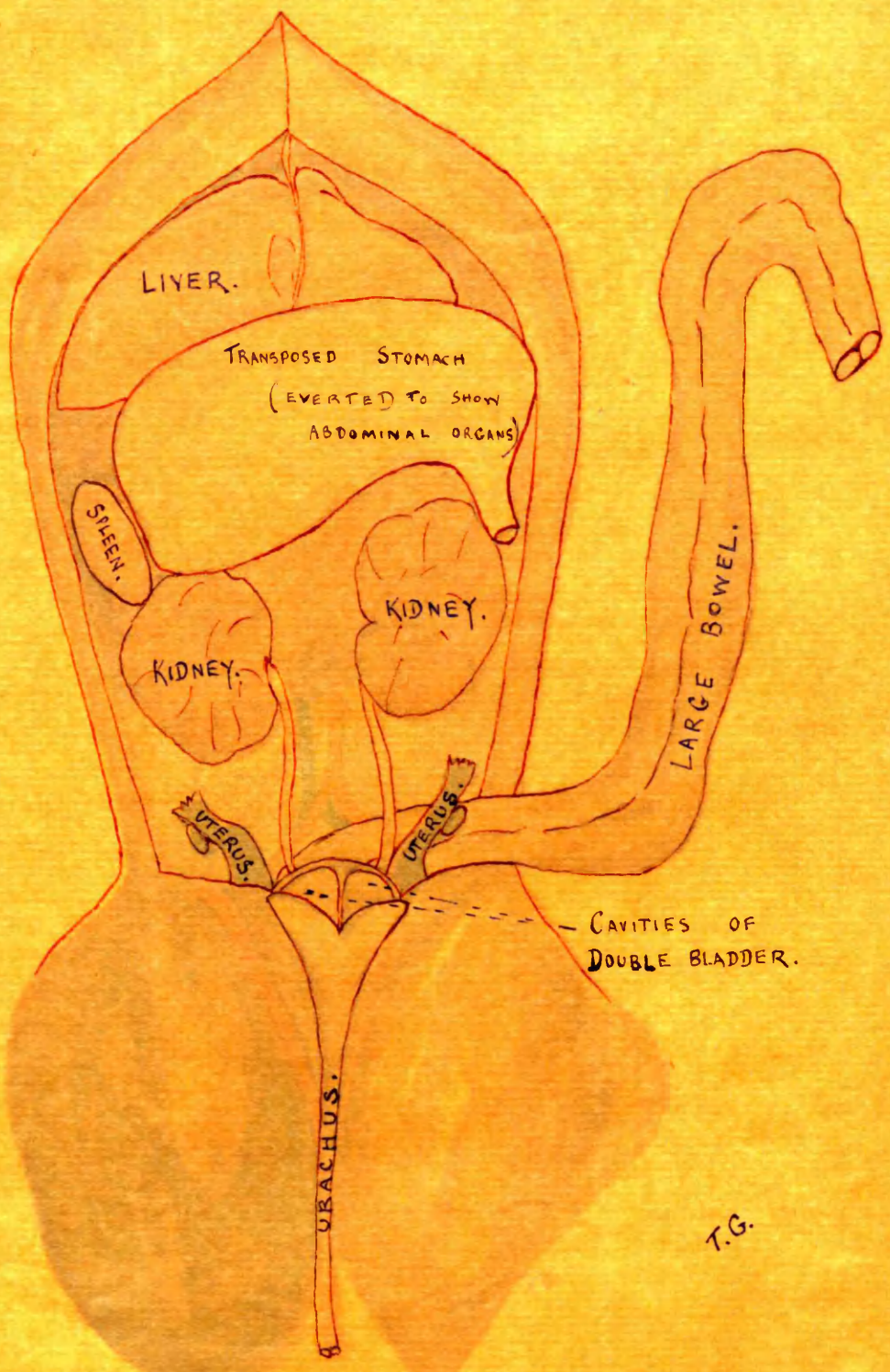
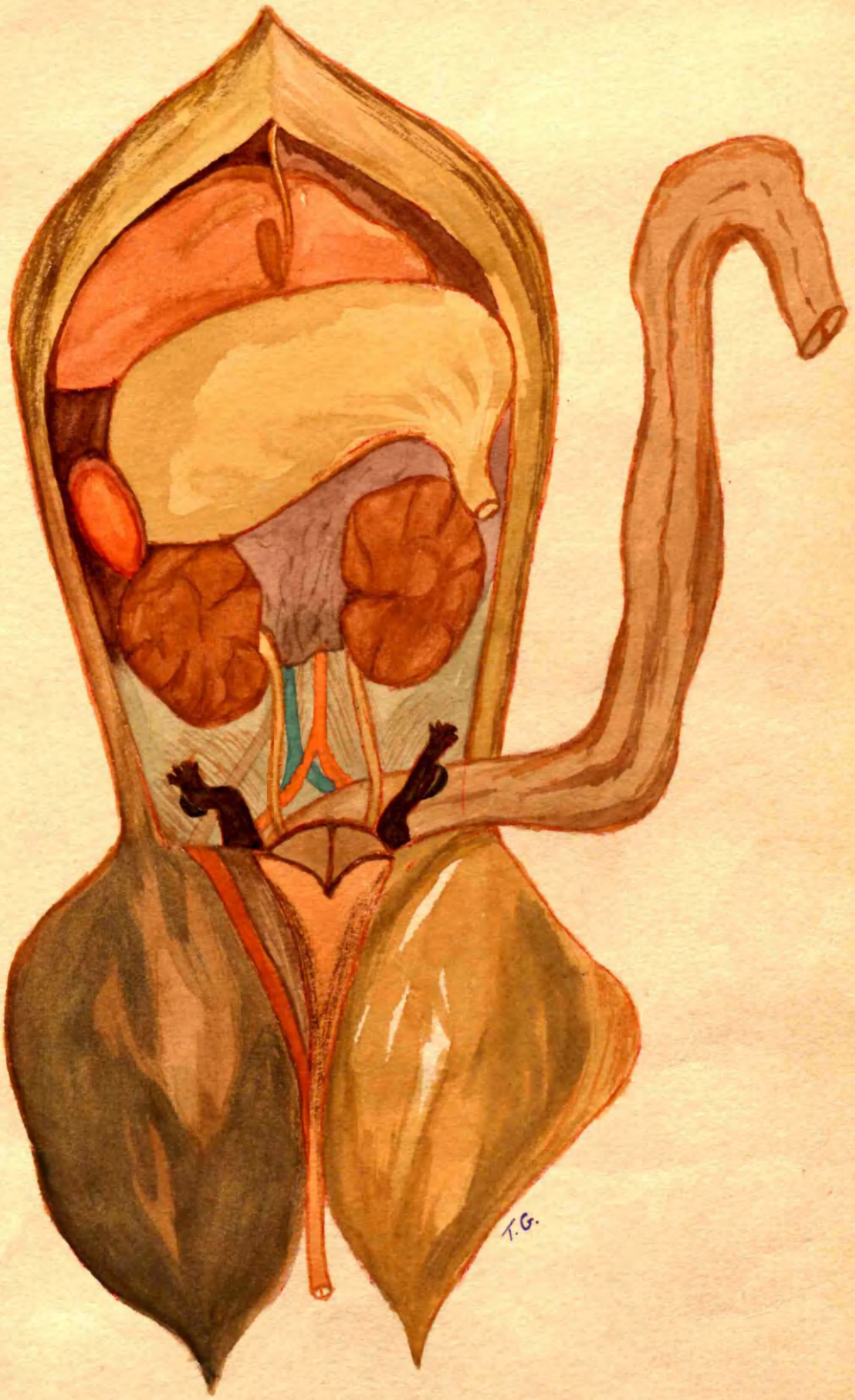


FIG. 8 - AUTHOR'S IMPRESSION OF THE ABDOMINAL AND PELVIC ORGANS AT AUTOPSY.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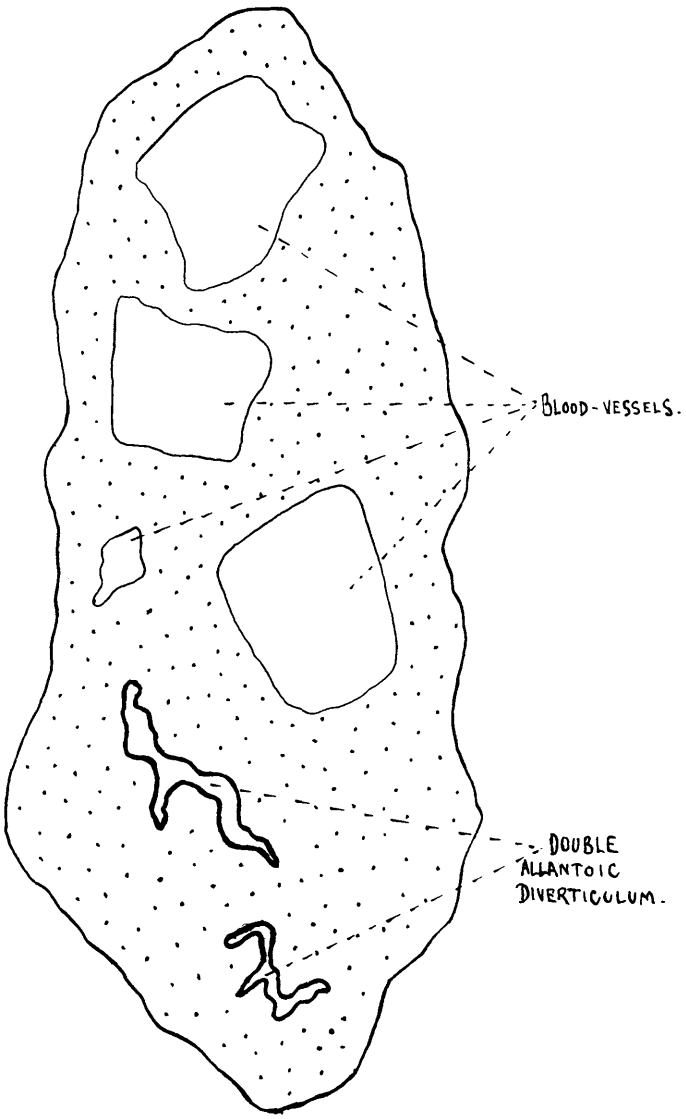


ig. 9. Diagrammatic Drawing of cross section of Urachus, showing Double Allantoic Diverticulum.

Both bladders had a urethral orifice from which ran a urethra. Both urethrae were patent in their whole extent.

There were two uteri situated widely apart, to each of which was attached a single tube and ovary. There were two vaginae. A single ureter descended on each side from a normal kidney. The ureters entered their corresponding bladders.

The condition of the bowel was remarkable. The ileum at a point six inches from its lower end showed on its surface a sulcus, which was the beginning of a bifurcation. The ileum divided at a point five inches from its termin-:ation into two distinct and separate ilea, which passed on into two distinct caeca. To each caecum there was attached a normal appendix/

appendix. (Fig. 10, p. 65). The caeca continued into two ascending bowels, which seemed to fuse into one bowel but examina-:tion revealed the interesting fact that they lay in apposition in a single serous layer and really ran like the barrels of a double-:barrelled gun as two separate large bowels to again bifurcate just below the brim of the pelvis to form two diverging recta and each rectum ran downwards to terminate at its corresponding anus.

The oesophagus came through the diaphragm in the middle line and the stomach and spleen were completely transposed. Behind the stomach lay the pancreas, which consisted of two separate parts. The liver was normal in position and showed an over-developed caudate lobe/

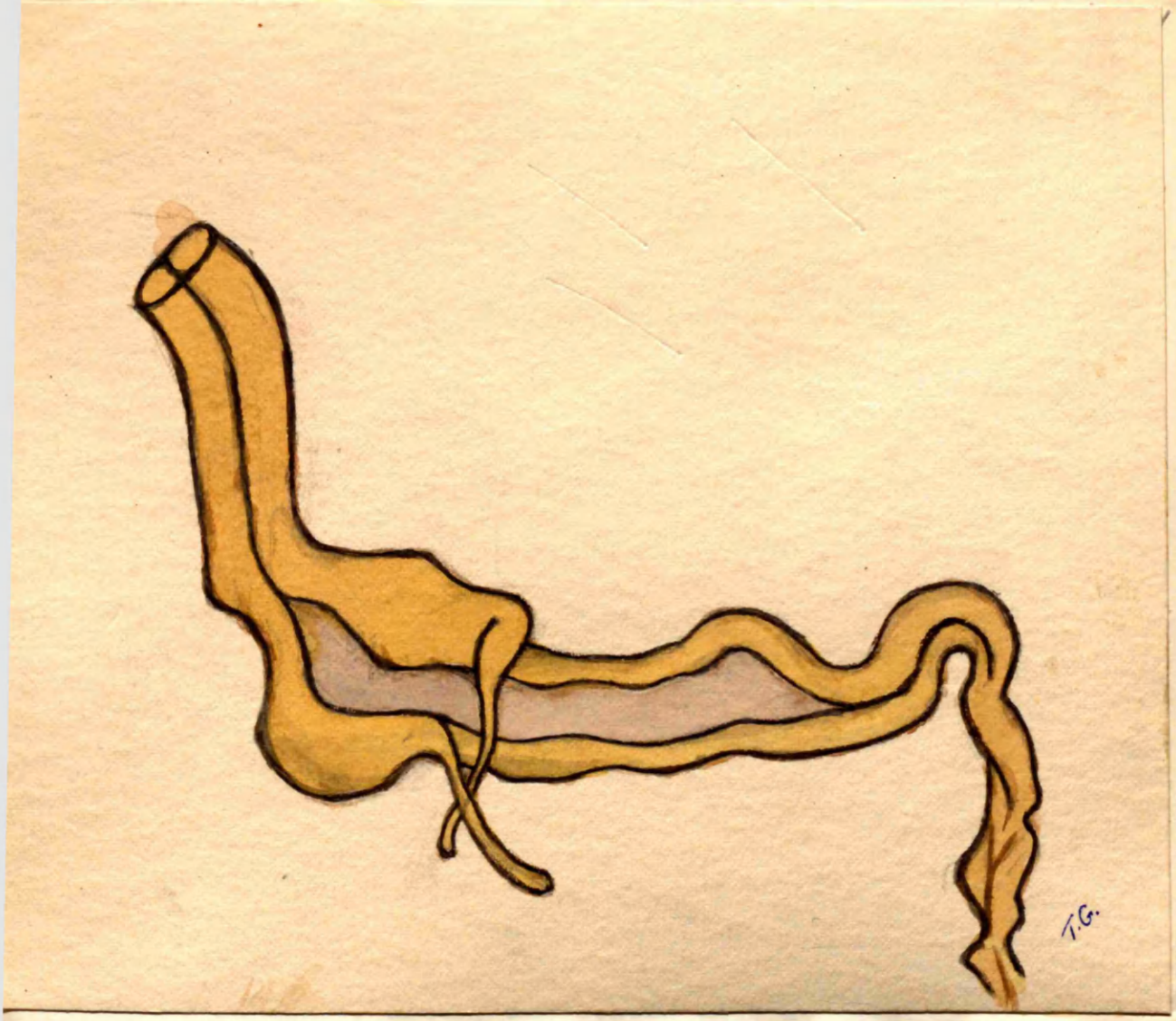


Fig. 10. Duplication of Terminal End of Ileum, Caecum and Appendix.

lobe.

Examination of the thorax showed the lungs and heart in normal position. The heart had, however, a well-marked defect in the inter-auricular septum and there was congenital atresia of the pulmonary artery. This had been the diagnosis clinically of and was the primary cause of death. It is interesting to note here that McCrae (34) of McGill University, Montreal, in 1913 record-:ed a case of congenital atresia of the pulmonary artery and stated that at that time only two cases were traceable in the literature available where a defect of this kind was combined with transposition of the viscera.

The meningocele was of large size with a thick/

thick wall. On incision it was seen to be bilocular. The spinal cord was prolapsed into it and nerve fibres could be seen distinctly radiating on its inner surface. There was irregular formation in the vertebral column.

Cleft palate was present.

FAMILY HISTORY.

The parents are young and are both members of large families. The mother's age is twenty-seven and the father's twenty-eight. The mother is the eighth member of a family of twelve and the father is the third member of a family of ten, in which twins have occurred twice. Blood taken from both parents for a Wassermann test showed a negative result. The father is healthy but the mother suffers from hypo-pituitarism. The twinning in the same generation on the paternal side, as can be seen from the family tree, has appeared with remarkable frequency. In all, there are on the paternal side nine cases of twin births and one of triplets but the most striking and impressive feature is the number/

number of twins of the same sex. A sister of the father is mentally defective. Cleft palate occurs in three generations. (Fig. 11, Diagram of Family Tree, p. 120).

C.

D I S C U S S I O N.

EMBRYOLOGY.

The study of the mode of origin of any abnormality arising in embryonic life is necessarily based upon knowledge of the normal embryology of the region involved. Before attempting, therefore, a solution of the abnormalities which have been described, one must consider:-

1. The early differentiation of the Embryonic Area.
2. The development of the Posterior Aspect of the Yolk Sac.
3. The development of the Uro-genital organs.

The early differentiation of the Embryonic Area.

The earliest stages of human development, as observed in Miller's embryo, is represented by/

by a blastocyst containing a solid mass of cells, the embryonic rudiment, in a matrix of delicate cellular structure. This jelly-:like matrix containing delicate cells, Bryce (35) says, represents the earliest stages yet known of the extra-embryonic mesoderm and "the generally accepted hypothesis regarding its origin in the human embryo is that it is budded off from the formative cell mass simultaneously with, or very shortly after, the differentiation of ectoderm and entoderm". Presumably this takes place while the cavity of the blastocyst is still quite small. The primitive mesoderm according to Bryce (36) is primarily an angioblastic tissue "and the generally accepted hypothesis regarding its origin is that it is budded off from the formative/

formative cell-mass simultaneously with, or very shortly after, the differentiation of ectoderm and entoderm".

The solid embryonic rudiment becomes converted into ectodermic and entodermic vesicles through the formation of cavities in the solid mass of cells. The embryonic area, that area from which the embryo proper is developed, is the region where the ecto-:dermal and entodermal vesicles lie in con-:tact. In its early stages the ectodermal is the larger vesicle but later the entodermic vesicle increases rapidly and becomes the larger of the two vesicles.

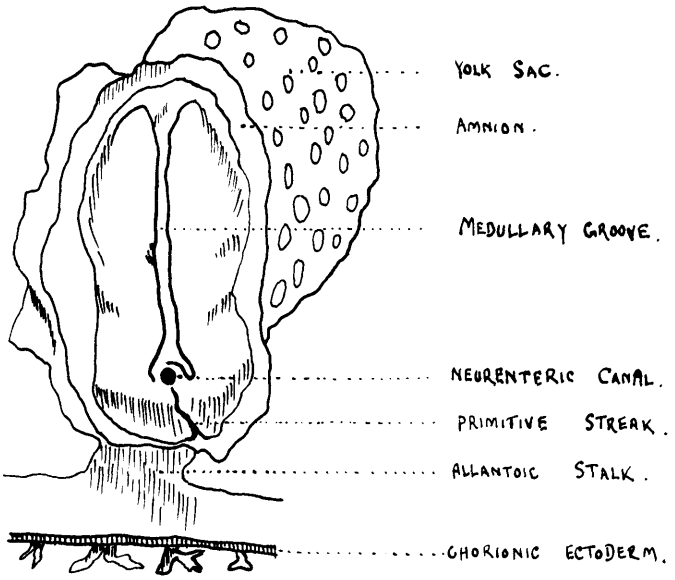
The embryonic area, at first rounded in shape, becomes oval and develops a wide anterior and a narrow posterior aspect. A thickened/

thickened ridge of cells, called the Primitive Streak, appears on the posterior portion and in the Primitive Streak a depression appears called the Primitive Groove. Probably a close connection exists between ectoderm and entoderm in the middle line. From the lateral margins and posterior end of the Primitive Streak is formed the Primitive Streak Meso-:derm. The Primitive Streak Mesoderm, un-:like the primitive mesoderm, is therefore relatively late in appearing and in its beginning is of small amount.

The medullary folds with the interven-:ing medullary groove appear in the ectoderm lying in front of the primitive streak. These folds, which constitute the anlagen of the nervous system, fold over to fuse and

form/

form the neural canal. At the anterior end of the primitive groove the neurenteric canal develops and forms a communication with the entodermal vesicle. By the fold-:ing over of the caudal ends of the medullary folds the neurenteric canal becomes included within the neural canal. "The neurenteric canal marks the part at which the entodermal vesicle (that part which becomes the hind gut) opens on the ectoderm". (Keith (37).) The neurenteric canal connects neural canal and gut and opens into the posterior aspect of the notochord. From the neurenteric canal the primitive streak and groove run backwards in the middle dorsal line to end on the allantoic stalk (Fig. 12, A. and B, p. 76), and it is this part that gives rise to/



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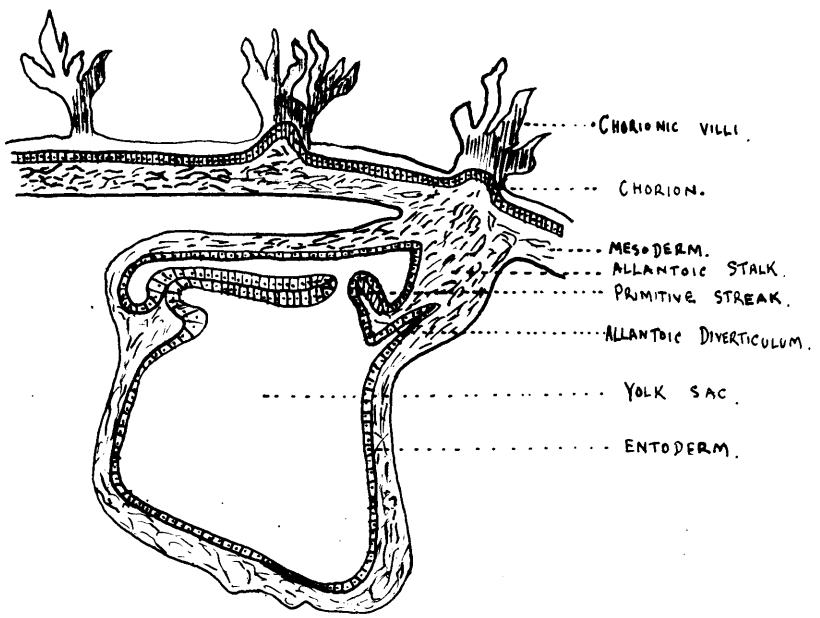

Fig. 12.A. Reconstruction of Embryo Gle (Graf Spee).

B. Mesial longitudinal section through Embryo Gle (Graf Spee).

to the median portion of the body and forms the perineal section, in which the openings of the rectum and urogenital passage are formed.

The differentiation of the cells of the embryo results in the formation of the three germ layers, ectoderm, entoderm and mesoderm, from each of which certain types of tissues or organs are developed.

The Development of the Posterior Aspect of the Yolk Sac.

When the embryonic area is folded into the form of the embryo the entodermal vesicle consists of three parts:-

1. A part enclosed in the embryo which forms the primitive entodermal alimentary canal.
2. A part lying outside of the embryo.
3. A third part connecting these two called the/

the vitello-intestinal duct.

The posterior diverticulum of the intra-:embryonic part of the yolk sac forms the hind-gut and the allantoic diverticulum. The anterior end of the hind-gut forms the post-:erior limb of a U-shaped tube, which is con-:nected with the yolk sac by the vitello-:intestinal duct. This caudal limb ultimately forms the terminal part of the ileum, caecum, vermiform appendix, ascending and transverse colon. As the tail of the embryo bends ventrally the hind gut becomes shut in and in that way becomes bounded in front by the posterior part of the primitive streak. (Fig. 13, p. 79.) The terminal part of this enclosed gut expands into the Entodermal Cloaca, which receives:-

1./

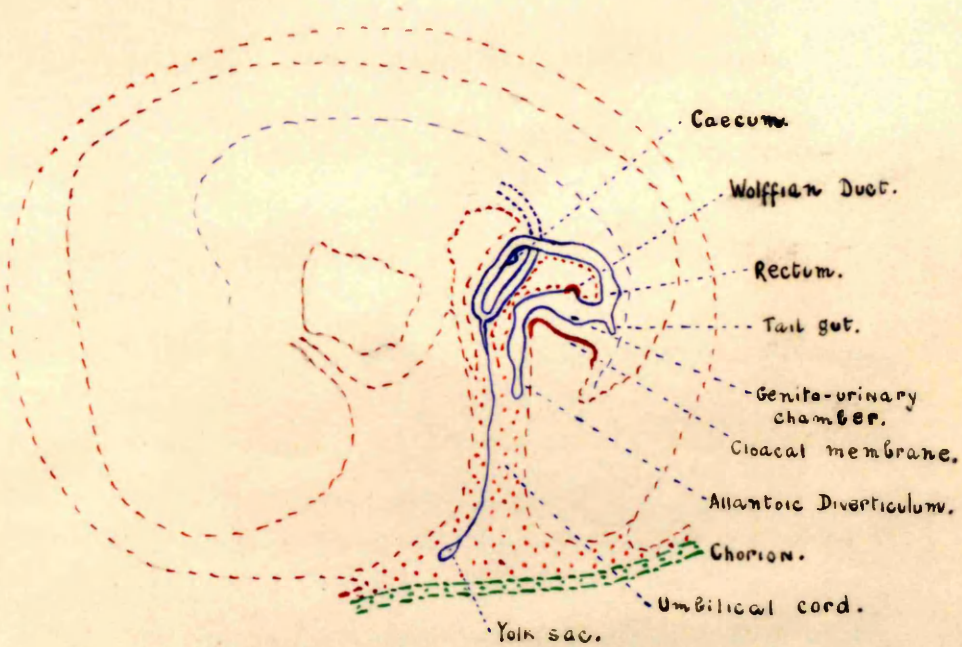

Fig. 13. Hind-Gut - before the separation of Entodermal Cloaca into Dorsal and Ventral parts.

1. The lower end of the alimentary canal.

2. The allantoic diverticulum.

3. The two ureters, which open laterally into the ventral side of the cloaca.

4. In the female the united Mullerian ducts, which end in the angle that joins the alimentary canal and allantoic diverticulum.

The dorsal part of the cloaca and ali-:mentary canal become shut off from the ventral part. This ventral part with the allantoic diverticulum is divided into three:-

1. An anterior part, which is converted into the urachus and apical part of the bladder.

2. A middle portion which forms the greater part of the bladder.

3. A caudal portion, which in the female becomes/

becomes the urethra and the vestibule of the vagina, and in the male the pro-:static and membranous part of the urethra. (Fig. 14, p. 82.)

Along the line of the primitive streak on the ventral aspect of the cloaca a depres-:sion appears in the ectoderm. This is the proctodaeum. In the floor of this depression the ectoderm and entoderm come in contact and form the Cloacal Membrane. By rupture of the anal membrane and union with the cloacal mem-:brane, the anal canal and anus are formed.

From the hind gut, therefore, the follow-:ing are derived:- (Robinson, (38).)

1. The terminal portion of the ileum.
2. The whole of the large intestine, except the anal canal and anus.

3/

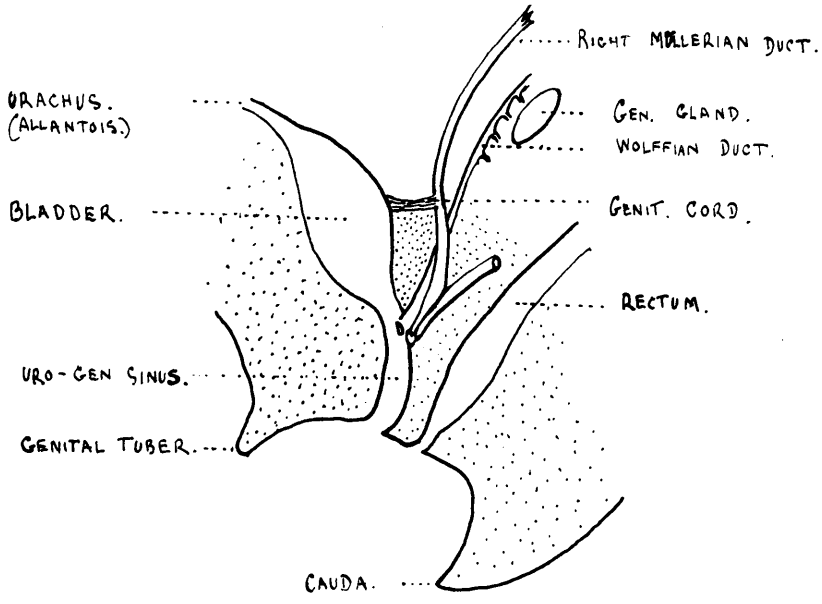

Fig. 14. Hind-Gut - after the separation of Cloaca into Dorsal and Ventral parts.

3. The urachus, bladder, urethra, the vestibule of the vagina in the female and the greater part of the urethra in the male.

The Development of the Urogenital Organs.

The urogenital organs arise from the allantoic diverticulum, anterior part of the cloaca, Wolffian ducts and Müllerian ducts. The part played by the allantoic diverticulum and anterior part of cloaca has been considered.

The Wolffian duct serves as the canal for the primitive secretory organs, which develop in connection with its anterior part. In both sexes the ureter arises as an outgrowth from a Wolffian duct. The Müllerian ducts follow in development. The tail end of the Müllerian duct descends with the Wolffian duct/

duct in the same peritoneal fold called the genital cord. Within this cord the uterus and vagina are formed from a fusion of the Müllerian ducts. In the lowest mammals and in the human foetus at the second month the Müllerian ducts remain separate and open in the cloaca. (Fig. 15, p. 85). The process of fusion begins in the third month and by gradual disappearance of the septum the uterus and vagina are formed. In the forma-:tion of the vagina according to Berry Hart (39) the mesoderm of the lower end of the urogenital cord grows rapidly and pushes the urogenital sinus downwards. The epithelial lining of the Müllerian ducts forms solid cords lying side by side which extend into the urogenital cord, fuse together and then break/

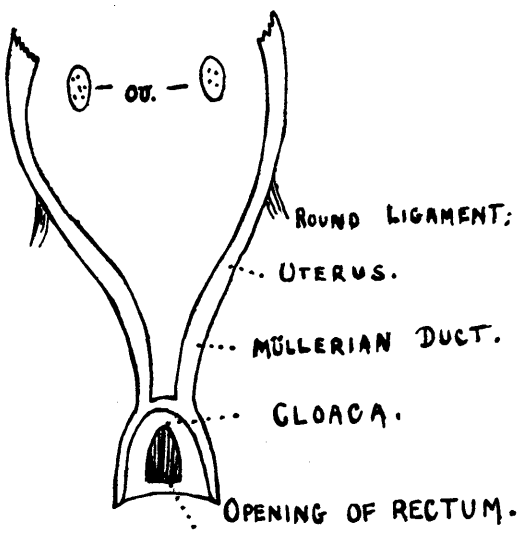

Fig. 15. Form of Müllerian Ducts seen in lowest mammals and in the second month human foetus. (after Keith).

break down to form the vagina, which opens externally behind the urethra in the uro-:genital cleft in the ectodermal cloacal fossa. This slit-like opening is similar in the male and female until the third month. At its anterior extremity appears the Genital Eminence, which becomes the clitoris or penis according to sex and there arise on each side of it the Labio-scrotal folds. After the third month the labio-scrotal folds develop into the Labia Majora in the female and the Scrotum in the male.

In the case of the male the inner genital folds, which correspond to the fourchette and labia minora, grow forwards and close the urogenital sinus. In this way the floor of the penile urethra is laid down.

The/

The urogenital sinus extends forwards and forms its roof, which corresponds to the vestibule of the female. The last part of the urethra to be formed is that part which traverses the glans. A solid rod of ecto-:derm cells is seen at this stage running backwards in the glans to meet the penile urethra. Just before birth this is con-:verted into a canal and the urethra is completed. As has been stated above, the prostatic and membranous parts of the urethra are developed from the urogenital sinus. (Fig. 16, p. 88.)

The scrotum, developed from the labio-:scrotal folds, is formed during the fourth month and the median raphé is a relict of their union.

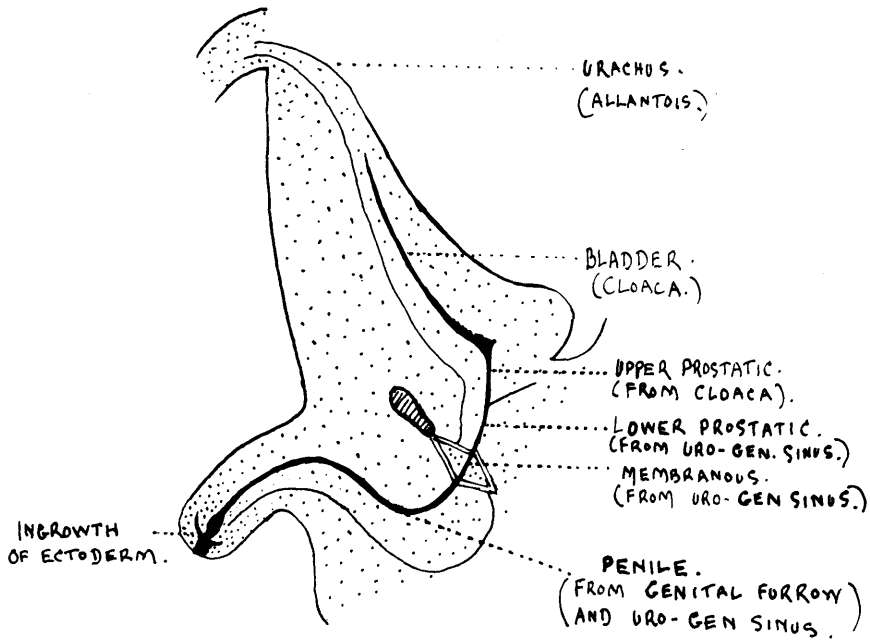

Fig. 16. A section of the male bladder and urethra at birth, showing the structures derived from the intra-abdominal part of the Allantois and from the Cloaca.

EXPLANATION OF THE ABNORMALITIES.

The theories that have been postulated for many years for the mode of origin of double malformations have been numerous and in most instances based on the principle of fusion or fission or a combination of these. The fusion theory presupposed a second foetus, an extreme view that could not account for the exact relationship of the two parts that are so symmetrical as in true double malformations, or suitably explain the cases of minor duplicity.

The fission theory accounted for double malformations by a division of the formative material normally belonging to one embryo. Windle (40), who supported this theory, attempted an explanation of the cause in attributing/

attributing this division to an excess of the germ plasm, the excess being due to (a) faulty segmentation of the polar bodies, (b) faulty formation of the spermatozoon or (c) poly-:spermy. He was of the opinion that, as the result of this excess of germ plasm, two grow-:ing points appeared in the ovum. These grow-:ing points were completely or incompletely double. If completely double they gave rise to identical twins. If incompletely double they gave rise to double malformations.

In 1904 Wilder (41) and (42) advocated a theory differing little from Windle's theory. He saw as the mode of origin of double mal-:formations a partial fission of the first two blastomeres, the extent of the fission determining the nature of the double malformation.

The/

The researches of Patterson (43) on the mammalian egg of the Texas Armadillo regard-:ing the question of polyembryony which in-:variably occurred in this animal formed a worthy contribution to the subject. The question of polyembryony is closely assoc-:iated with the interesting problem of the origin of identical twins and double malforma-:tions. This animal produces constantly a litter of four, all males or all females. In studying the early embryonic stages Patterson found that a budding process appear-:ed in the single blastocyst. Two primary buds arose from the ectodermal vesicle in the form of two diverticula. When these were formed, the two secondary buds arose and thus four embryos were established within the single/

single chorion in two pairs. Patterson, referring to his conception of the mode of origin of polyembryony, says "I am encouraged to make the suggestion because of the recent discoveries in the human ovum, (e.g., Bryce - Teacher ovum, 1908), in which the condition of the ectodermal vesicle is shown to be such as to require no great stretch of the imagina-:tion to picture how diverticula might arise from it and thus initiate the development of two or more embryos. Nor is there any greater mental strain in accounting for the origin of composite monsters in this way than is required in the hypothetical juggling of blastomeres to account for the various relationships and positions assumed between the components of these monsters". It was discovered that the blastocyst/

blastocyst underwent a resting process in the uterus before becoming embedded and it was thought that the resulting arrested development was responsible for the alteration of structure in the ovum, the alteration occurring just previous to the primitive streak formation. A similar resting process, however, occurs in the deer without resulting polyembryony.

This would seem to prove that the predominant factor in producing polyembryony or the allied phenomenon, double malformation, is the inherent quality of the fertilised ovum.

Within recent years our knowledge of the mode of origin and cause of double malformations has been greatly increased as the result of advances made in Experimental Embryology.

One/

One of the most stiking of its early results was the production of twin individuals from a single egg by separating the two primary blastomeres, each blastomere pursuing an independent development and giving rise to a complete individual. The conclusion was readily formed that the occurrence of iden-:tical twins resulted from the separation of the primary blastomeres, due either to mechan-:ical interference or chemical cause. But when the eggs of different species were sub-:jected to this experiment the end result was not two complete but only two partial indiv-:iduals. The theory was further disproved by Stockard (44), who showed that when the ovum was interfered with mechanically in the late cleavage stages identical twins and double malformations/

malformations could be produced.

Stockard's experiments furnished re-
:markable results. He showed that by arresting development of the embryo in any one of the following ways (a) by lowering the temperature of the medium in which the embryos were developing, (b) by diminishing the supply of oxygen to the embryos or (c) by treating the medium with chemicals, he could produce any known deformity. This re-
:sult could be obtained successfully only if the embryos were treated before the end of gastrulation. The developmental anomaly depended on the time of interruption. "An arrest of development before gastrulation" he says "may cause a blastoderm to form two embryonic processes which may later develop into/

into a double monster. The degree of duplicity in double individuals depends upon the original distance apart of the embryonic buds on the blastoderm".

A few years later, 1923, Riddle (45) extended the field of experiment and obtained the same results by accelerating development by subjecting the fertilised ovum to the effect of increased temperature. He maintained that identical twins or double malformations were the result of alteration of developmental rate.

In 1925 Marie A. Henrichs (46) treated the eggs of fish with ultra-violet radiation at varying intervals after fertilisation and produced the same types of deformities that had been obtained by subjecting the ovum to heat/

heat, cold or chemical solutions. From her experiments she found that anterior duplicity followed the radiation at or before the forma-:tion of the two primary blastomeres. As time went on it became increasingly difficult to disturb normal development though "post-:erior duplicity appeared more regularly when the egg was radiated at the time when the posterior region of the embryonic axis was apparently a region of relatively high sus-:ceptibility".

Normal development depends therefore on a stable environment. Experimental Embry-:ology has proved that if this stability is affected by influences, even widely differ-:ent in nature, modification can take place in development and if the developing embryo

is/

is thus affected in its early stages, the resulting modification may result in a double malformation.

When Gemmell and Paterson (30) described their classical case of posterior duplicity the factors they held responsible were:-

1. Absence of the Allantois and consequent absence of the Allantoic diverticulum.
2. Traction of the Amnion.

According to their theory the bilateral bladders, owing to the absence of the allantoic diverticulum, developed from the ventral part of the Internal Cloaca. This was favoured by the traction of the Amnion which was held also to be the cause of the separate connection of the Müllerian Ducts with the

External/

External Cloaca. "It is probable", they say, "that the events which occurred to pro-:duce the anomaly happened in early embyonic life and that the formation of two vulvae was a secondary event - resulting from the division of the External Cloaca into two parts and the concomitant sub-division of the phallic papilla". The authors regretted the absence of any informa-:tion at the time of the patient's birth and no information of her family history was given. This explanation of the abnormalities can be set aside here. In both of my cases the placentas, membranes and umbilical cords were normal as far as the naked eye could tell and the umbilicus was normal in size and position. Moreover, in the cases of posterior duplicity recorded in literature no abnormality is described/

described in the position of the umbilicus or in the formation of the placenta or membranes.

It has long been debated whether or not the Amnion plays any part in the production of malformations. One can see how, through the effects of pressure or adhesions, the Amnion might modify the growth of a normal structure and in that way be responsible for a congenital abnormality but it is difficult to believe that it could play an important part in a developmental abnormality of this kind.

D. Berry Hart, Edinburgh, (47) dis - :cussed Gemmell and Paterson's case at the time it was recorded and differed from the authors in their interpretation of the condition/

condition. He pointed out that the bladder, except at its apex, is developed from the ventral part of the Entodermal Cloaca and not from the Allantois. Hart admitted the great difficulty of interpreting the double vulva, double bladder and double urethra. He sup-:posed a doubling of the determinants for the Entodermal Cloaca and external genitals. The doubling of the former would necessarily re-:sult in a double bladder, urethra and rectum.

In my cases the outstanding features are:-

1. In the male: the complete duplication of external genitalia in an individual other-:wise normal.

2. In the female: the complete duplication of external genitalia, duplication of urinary and genital tracts, bladder and uterus/

uterus, duplication of bowel from the terminal end of ileum to anus and double anus: transposition of viscera.

The two cases represent different degrees of posterior duplicity. In the female case, as the result of a budding or branching process of the cells of the posterior end of the Embryonic Area that give rise to the Primitive Streak, a double Primitive Streak appears. The posterior ends of this double Primitive Streak converge on the narrower posterior extremity of the embryo.

We can understand better this modification of growth when we consider parallel instances in plant growth. In the growth of the trunk of a tree which is normally a single/

single stalk we sometimes see a budding pro-
:cess which gives rise to a forked growth.
A similar modification takes place sometimes
in the root of a plant. One commonly en-
:counters a forked root in a carrot. There
is of course this difference that, whereas
in a plant the growing points are active
during the whole of its life, this activity
in the mammal, in whom we can consider an
anterior and posterior growing point, ceases
on the formation of the medullary groove and
therefore the modification of growth at a
growing point can take place only in the very
early life of the embryo.

The neurenteric canal connects the en-
:toderm of the hind gut with the surface
ectoderm and also opens into the posterior

end/

end of the canal of the notochord. "As it opens at the anterior end of the Primitive Streak, which afterwards is included in the posterior end of the neural groove, such a canal, if it persisted", Keith (48) says "might be expected to perforate the coccyx". The posterior end of the Primitive Streak originally ends on the Allantoic Stalk.

In the median line, therefore, extend-:ing from the neurenteric canal to the umbil-:icus the anus, perineal depression and genital parts are formed. If in our female case we consider that a branching process has taken place at the anterior of end of the Primitive Streak, we have the condition of affairs laid down as represented diagramatically in Fig. 17, p. 105, which would give rise to double vestigial/

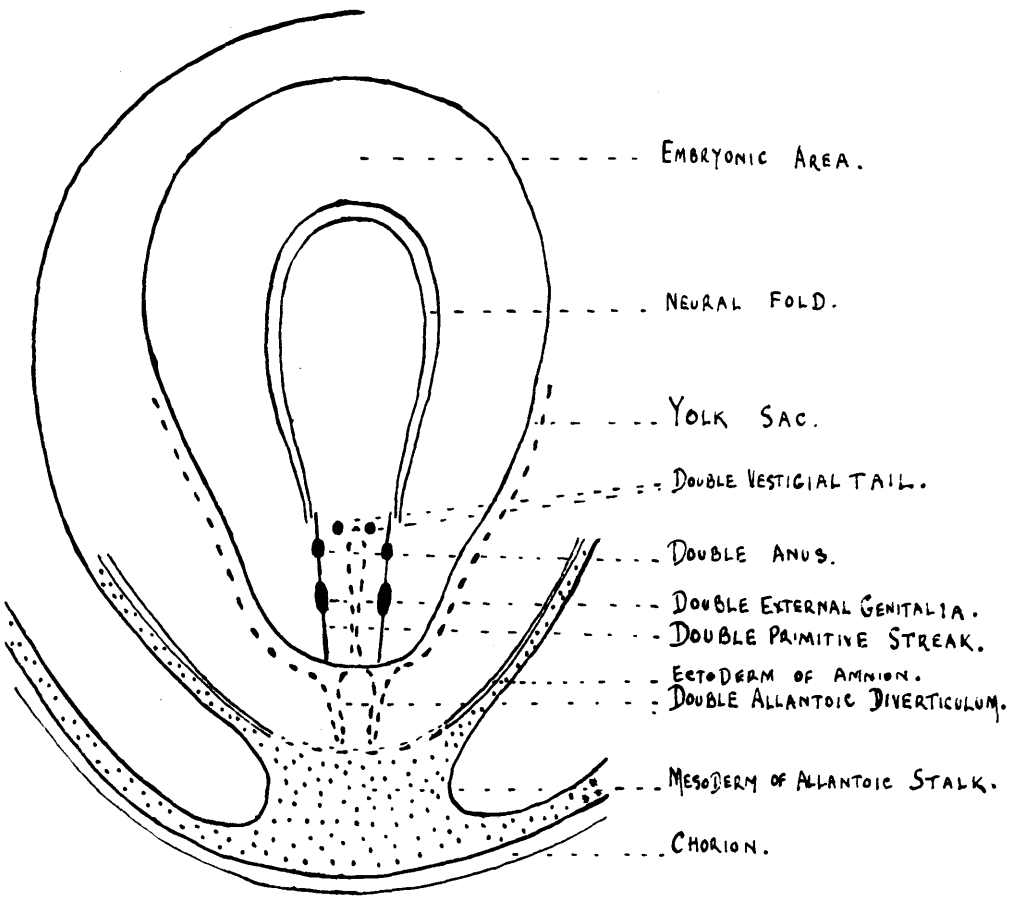

Fig. 17. **Diagram of Dorsal Surface of Embryonic Area, showing how a Double Primitive Streak and Structures seen in the Author's Female Case might arise.**

vestigial tail, double anus and double ex-
:ternal genitalia.

In the female case the problem of great difficulty is to find an explanation for the wide-spread duplication of bowel, the duplication being wholly caudal to the vitello-
:intestinal duct.

When the branching took place at the Anterior end of the Primitive Streak the same process has affected the underlying entoderm, which is the entoderm of the hind gut. This has resulted in the formation of two diverti-
:cula in the posterior aspect of the Yolk Sac and has given rise to a double hind-gut with double allantoic diverticulum. There has thus arisen a duplication of the terminal end of ileum, caecum, appendix, whole of large

bowel/

bowel and cloaca, the double ventral part of which with double allantoic diverticulum has formed double urachus, double bladder and double urethra. The urachus, as has been stated above, was seen by microscopic exam-:ination to be double, offering proof that there must have been present in the early embryo a double allantoic diverticulum.

The Müllerian ducts have descended to enter separately their corresponding Ento-:dermal Cloaca situated on either side of the middle line and link up with the lower third of either vagina, which is developed from either urogenital sinus.

The presence of transposition of the stomach and spleen with extensive duplication in the female case is interesting. Morril (49) has/

has found in double fish a transposition of organs occurring far more frequently than in any group of single vertebrate individuals. It would seem that transposition of viscera was associated with the double condition. We know that transposition of viscera has been known to exist in man in a single in-:dividual. It is not unlikely that a number of these cases would prove, if complete exam-:ination could be carried out, to have some form of twin manifestation and be really in-:complete twins.

In the male case the branching process has taken place at a point immediately in front of the position in the median line where the genital part normally appeared and there-:fore the double malformation was confined to the/

the external genitalia. There has thus been a doubling of the anlagen of the external genitalia. Two genital eminences have been formed. The right has developed in a normal manner but the left in the absence of a connection with the urinary tract has given rise to penis, scrotum and urethra only partially patent.

In looking for the cause of the malformations in these two cases we have to consider two striking facts in the family history, the presence of endocrine disturbance and the remarkable history of twinning on both the maternal and paternal sides.

We have already seen that it has been established by experiment on lower animals that abnormal development can result from the injurious/

injurious effects of chemicals on the fertil-
:ised ovum and it has been pointed out that
poisoning by such a chemical as lead has given
rise in mammals to a high percentage of mon-
:strosities. This would seem to point de-
:finitely to the influence of a poison in
circulation having an effect on the germ cell
of the parent and being a factor in the occur-
:rence of congenital malformations. There
is no reason why alteration in the constitu-
:tion of the blood or lymph should not affect
the germ cells and no more important agent
could be held responsible for that effect
than the altered secretion of the internal
glands although, of course, direct evidence
of the operation of such conditions is im-
:possible to obtain. We have noticed in
the/

the above two cases the presence in the relatives of hypo-pituitarism and cretinism. Whether we accept the evidence or not we must admit the likelihood of the serious effect on the germ cell of disturbed secretion of the internal glands and the possible consequent occurrence in the offspring of some form of deformity.

The question of heredity in the causa-:tion of malformations is of great importance, and though its transmission is as yet a pro-:blem unexplained, no apology is made for alluding to this interesting question. It is well recognised that a malformation is often traceable through many generations. In the family tree depicted later, the presence of cleft palate, for example, is seen in three

out/

out of five generations, but the occurrence of plural births is unique. It is well nigh conclusive proof that the twinning process is truly inherited. Certain mothers produce eggs which have a strong potentiality to-:wards twin formation and the family tree in my cases would afford proof that such property may be transmitted or inherited by the daughters and by the sons. A study of this family tree shows that the male can transmit the twinning tendency equally as the female. Moreover, in certain families the occurrence of twins is of much higher frequency than in other families. This property may be cited as an example of a dominant property possessed by the germoplasm of a particular strain, but the occurrence of two cases of duplicated external/

external genitalia which after consideration I regard as incomplete twins (and I have Wilder's support for this conclusion), is an example of the element of degeneration that is sometimes seen in reproduction. Might not the element of degeneration result, as was possible in these two cases, from the effect of endocrine imbalance, which in the light of recent knowledge we know to be a prominent factor in nutritive disturbances and therefore might alter the composition of the germ cell of the parent or the nutritive conditions in which development takes place?

Never previously has there been recorded a family tree showing two cases of a rare double malformation and one is forced to conclude that the study of the parent germ cell/

cell has not found a prominent enough place in the theories propounded. The cause of double malformations may lie, therefore, not only in the unusual environment but also in a particular type of germ cell and influence the course of development from the very be-:ginning. If this hypothesis, which seems reasonable, is true, the germ cell which gives rise to double malformations differs from normal as the modified development which arises differs from normal.

In the preceding discussion one is only too conscious of the problems and difficulties that arise, when an attempt is made to correlate instances of duplication in mammals with the striking results obtained from experimental work on lower animals. Although a definite

cause/

cause may produce a double malformation by experiment, the natural cause of a double malformation may not be found. From many of these results we cannot justifiably reason, but on the other hand, as regards the effect of injurious agents on the ovum, we may learn much from experiments that may assist us some day in preventing harmful results on the embryo in the respect at least of minor deformities. As we have no field for experiment with mammals, as with the lower animals, the subject of the malformation must be thoroughly studied, subjected to minute dissection and even to microscopic investiga-:tion. Nor should we omit the fullest exam-:ination of the parents or fail to make an exhaustive search into the family history.

The/

The investigation of all these points is necessary if we would arrive at an accur-:ate knowledge not only of the mode of origin of double malformations but of the very obscure question of the cause.

SUMMARY.

1. Duplicated external genitalia in the male and female is a rare condition.

2. The life of the foetus is not affected by the presence of this malformation.

3. Many of the cases in the male are really bifid penis. The homologous condition in the female is excessively rare.

4. There is a tendency in both sexes to arrested development in the parts.

5. In the male two normal scrotal sacs are rare and in the female the labia majora, especially on the outer aspect, are usually better developed.

6. The rarest of all associated defects are double bladder, double anus and double bowel.

7/

7. Transposition of stomach and spleen has been met with in the female case of ex-:tensive duplication. Single individ-:uals in which transposition of viscera occurs may be incomplete twins.

8. Family history has been rarely investi-:gated in this interesting problem.

9. In the author's cases the presence of endocrine disturbance has been noted in near relatives but of more outstand-:ing interest in the family history is the frequent twinning on the maternal and paternal sides.

10. Duplication of external genitalia with associated duplication is possibly due to a branching process of the Primitive Streak.

11/

11. Normal development depends on a stable environment, the disturbance of which may result in double malformations.

12. The cause may lie not only in unusual environment but in a particular type of germ cell. This may influence the course of development from the beginning and control throughout the modified develop-:ment of altered structural details laid down in the ovum in its earliest stages.

13. Minute examination of the subject of double malformation, fullest examination of the parents and exhaustive search into the clinical and family history are necessary for the elucidation of the mode of origin and cause.

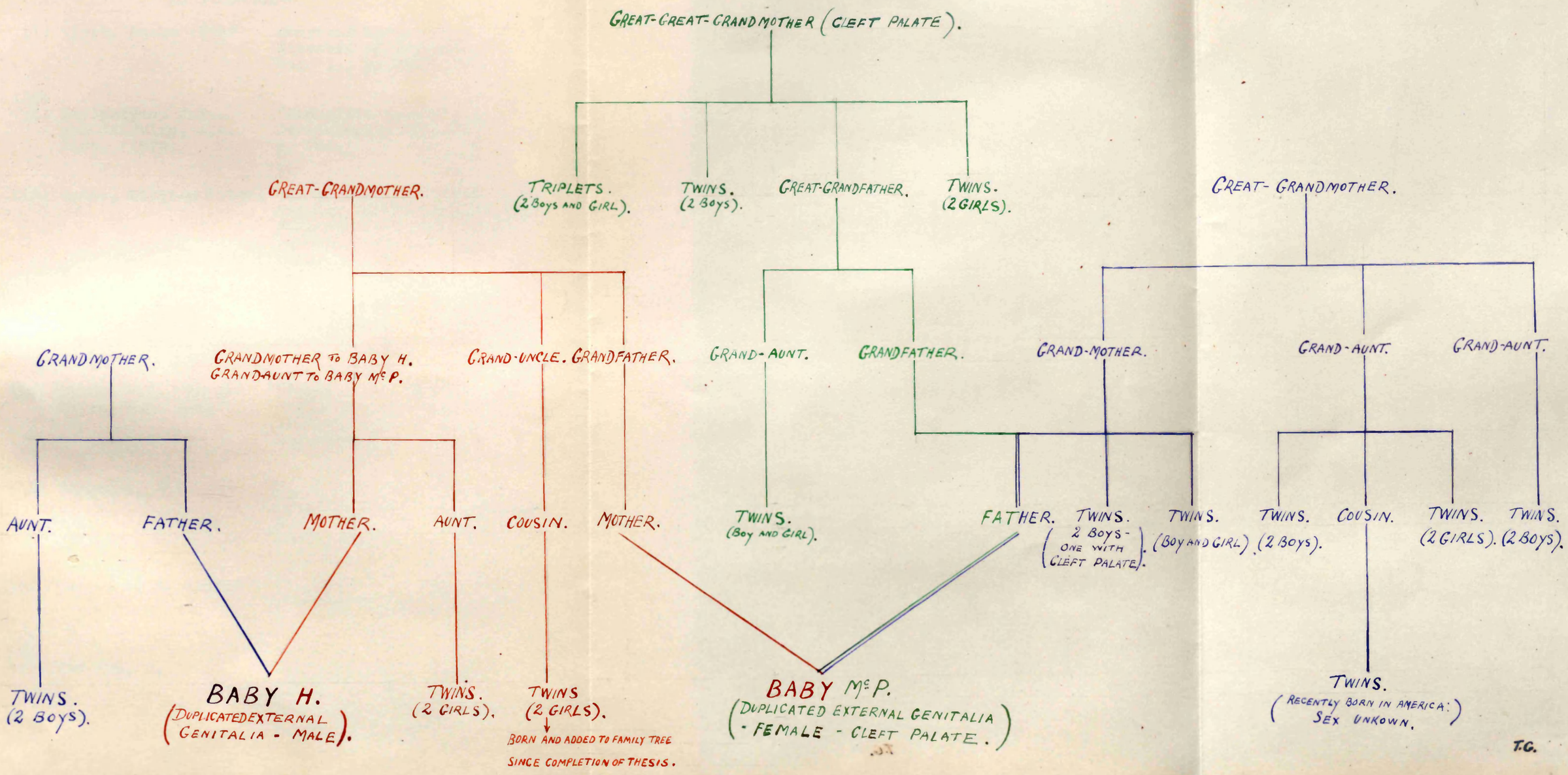

FIG. 11.- DIAGRAM OF FAMILY TREE SHOWING OCCURRENCE OF TWO RARE ABNORMALITIES AND REMARKABLE FREQUENCY OF TWINNING ON MATERNAL AND PATERNAL SIDES.

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