tation of the fundus (*uterus arcuatus*) the lack of fusion is merely suggested. From this condition all gradations are seen to the complete separation of the tubes (*uterus bipartitus*), at times involving the entire utero-vaginal tract (*uterus didelphys*).

The Müllerian duct of one side may undergo partial obliteration, occasioning the closure and disappearance of the corresponding portion of the doubled uterus, in which one horn may be represented by a solid cord (*uterus unicornis*). Unilateral arrest of development may also affect the Müllerian ducts as shown, when of slight degree, by the asymmetry and obliquity of the uterine fundus. Very rarely the fused ducts may remain so rudimentary that later they are represented by only a solid muscular nodule or cord.

Not infrequently the uterus retains the *infantile form* in consequence of failure of the body segment fully to develop, the cervical portion being unusually prominent as in early life.

The *vagina*, having similar relations to the Müllerian ducts, is subject to the same congenital duplicity as the uterus, although the septum is frequently partial and incomplete. One of the two canals may be partly obliterated by unilateral arrested development of the embryonal tubes. The vagina is very rarely entirely absent. The more usual malformations, however, include narrowing or closure, especially of the upper portion. Congenital vaginal stenosis is often associated with a similar condition, or obliteration, of the uterine cervical canal. Persistence of the infantile type of uterus is usually accompanied by stenosis of the vagina, although the vaginal defects may exist independently of abnormalities of the upper segments of the genital tract. The vagina may be partially represented by a solid cord. The *hymen* is seldom absent; the imperforate, annular, and cribriform types are some of the variations which this duplicature presents.

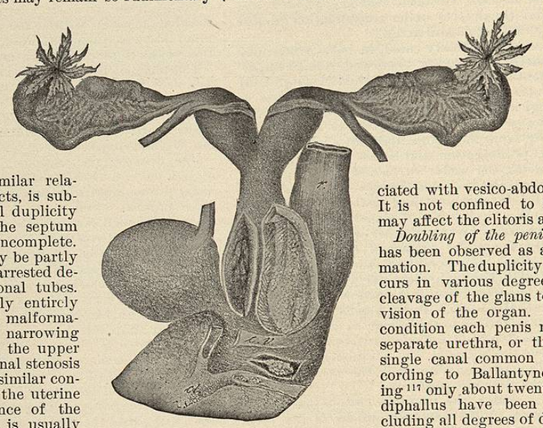

FIG. 4686.—Imperfect Fusion of Müllerian Ducts Resulting in Double Uterus and Vagina. (Schröder.)

MALFORMATIONS OF THE EXTERNAL SEXUAL ORGANS.—The external generative organs of both sexes are derived from three sets of structures—the *genital eminence*, the *genital ridges*, and the *genital folds*—which until the close of the third fœtal month are sexually undifferentiated. Elongation and fusion of these embryonal parts characterize the development of the male organs, while those of the female retain the original separated condition of their constituents.

The Male Organs.—The penis is evolved by the growth and extension of the unpaired genital eminence (from which are derived the corpora cavernosa), in conjunction with the elongation and fusion of the urethral ridges contributed by the walls of the urogenital sinus and the genital folds (from which are formed the corpus spongiosum and the urethra).

The penis may be entirely wanting, although this rare defect is usually associated with other malformations of the external genital organs. More frequently its development is rudimentary in consequence of which the organ remains small and clitoris-like. Arrested or faulty union of the folds, derived from the walls of the urogenital sinus, results in the production of a cleft along the under side of the penis (*hypospadias*) by which the urethra is opened, its termination lying at any point between the glands and the scrotum. When the arrest of development occurs at an early period, the cleft may be extensive and the urogenital sinus may remain entirely open. In such extreme cases the genital ridges, which normally unite to form the scrotum, may persist as separate folds embracing the hypospadial fissure. *Occlusion* and partial *obliteration of the urethra*, on the other hand, may occur in consequence of secondary excessive fusion of the uniting folds, or, when located at the glans, due to persistence if the epithelial urethral septum. Excessive size of the urethral crest surrounding the sinus pocularis has also been found responsible for closure of the urethra (Fuchs[116]). Enormous distention of the bladder and dilatation of the ureters are natural consequences of the obstruction of the urinary passageway. The urethra frequently communicates with the rectum in cases of atresia ani and, in the female, with the vagina when imperfect differentiation in the urogenital sinus exists.

The much rarer dorsal cleft (*epispadias*) is, as already pointed out above, usually associated with vesico-abdominal fissure. It is not confined to the male, but may affect the clitoris as well.

Doubling of the penis (or *clitoris*) has been observed as a rare malformation. The duplicity (*diphallus*) occurs in various degrees, from mere cleavage of the glans to complete division of the organ. In the latter condition each penis may enclose a separate urethra, or there may be a single canal common to both. According to Ballantyne and Skirving[117] only about twenty instances of diphallus have been recorded, including all degrees of duplicity.

Lange[118] has described an additional interesting case in which the two penises lay side by side, each connected with a scrotal sac containing a single testicle. There were two urethræ and two entirely distinct bladders, each with one ureter. The rectum was without anal opening and communicated with both urethræ. This condition, since the primary genital eminence is unpaired, probably depends upon profound early defects involving the cloaca and the urogenital sinus similar to those responsible for the production of epispadias and exstrophy of the bladder.

In rare instances the termination of the Wolffian ducts, later the seminal ducts, is continued during the development of the penis as an independent canal, the urethra being coincidently formed as a second one. In such cases the penis may contain two canals, the one for the products of the sexual glands, the other for the urine. Absence of the prepuce rarely occurs; oftener an abnormal shortening. On the other hand, not infrequently an unusual congenital redundancy exists.

The Female Organs.—In rare instances the external genital organs may be wanting, or so rudimentary that little differentiation into distinct parts takes place. On the other hand, congenital hypertrophy of the clitoris or labia is occasionally seen. Neugebauer[119] has reported the case of a young woman of twenty-two years who possessed a second clitoris, about one inch long, attached to the perineum. It presented the appearance of a diminutive penis, having perfect glans, corpora cavernosa, and partial prepuce, but was imperforate. There where no signs of testicles or other abnormal conditions. Chiarleoni[120] described a remarkable case, that of a child of about three years, in which there were two sets of external genitals, each possessing labia minora and vestibule; on the left side a clitoris was present, below which opened a minute orifice for urine and fæces. A similar aperture

opened into the right vestibule. An anus was wanting. The most conspicuous malformations are those arising from an attempt to follow the male type of development. In consequence of such deviation union of the genital folds and ridges takes place, resulting in closure of the vestibule and vagina. Since these malformations play an important part in the production of certain hermaphroditic forms, their consideration is appropriately included under the discussion of that subject.

HERMAPHRODITIC MALFORMATIONS.

Since the generative organs of the two sexes are differentiated from embryonal structures which are for a time common and neutral, it is not surprising that at times their development is irregular and results in the production of aberrant forms, in which certain characteristics of one sex are combined with those of the other. This applies especially to the generative ducts and the external organs, the sexual glands being very rarely of more than one type in the same individual.

In comparing the various types of sex duplicity it is necessary to bear in mind the salient features of the development of the sexual organs. The latter are produced by three independent but coincident processes: (*a*) the formation of the sexual glands; (*b*) the formation of the excretory ducts; (*c*) the formation of the external genitals.

The sexual gland develops from an anlage in the immediate vicinity of the Wolffian body; for a time indifferent, it later differentiates into testicle or ovary.

The Wolffian body consists essentially of the transverse tubules and their common canal, the Wolffian duct. An additional tube, the Müllerian duct, lies parallel to the former. The four tubes—the two Wolffian and two Müllerian ducts—are associated as the genital cord and open on the posterior wall of the urogenital sinus.

In the development of the male excretory passages, the Wolffian tubules and duct take the most important part, forming the vasa efferentia and coni vasculosa, and the tube of the epididymis and the vas deferens respectively. The Müllerian duct in the male is largely atrophic, remains of its upper extremity persisting as the unstalked hydatid connected with the globus major of the epididymis, while the fused lower ends of the two ducts are represented by the sinus pocularis, or uterus masculinus, on the posterior wall of the prostatic urethra. The sinus pocularis is, therefore, the homologue of the vagina and possibly the uterus. When the intermediate part of the Müllerian duct persists, it constitutes the duct of Rathke.

In the female type the active factors are the Müllerian ducts, the upper segments of which remain separate to become the oviducts, while the remaining parts unite and fuse into a common tube, which becomes the uterus and the vagina. The upper part of the Wolffian duct and some of the associated tubules persist as the epioöphoron, or the organ of Rosenmüller, the homologue of the globus major of the epididymis. The remaining parts of the Wolffian ducts ordinarily disappear, or at best remain to a limited extent as the tubules of Skene, which lie on either side of the vagina. Exceptionally the Wolffian duct persists and is then known as Gärtner's duct.

The external generative organs arise from the genital eminence, genital folds and genital ridges, in conjunction with the urogenital sinus. In the male the genital eminence enlarges to form the corpora cavernosa, the spongy body being formed by the extension of the walls of the urogenital sinus and the genital folds. The scrotum represents the fused and enlarged genital ridges, the raphe indicating the line of union. In the female the original separated relation of the genital ridges and folds persists, the former becoming the labia majora, the latter the nymphæ, while the genital eminence forms the clitoris. The urogenital sinus contributes the urethra and the vestibule.

The duplicity of sex in the same individual implied in the condition termed "hermaphroditism" is almost always only apparent and not real, since response to the crucial test, to be applied to every case by microscopical examination, as to the presence of both forms of sexual glands is rarely affirmative. Hermaphroditism is therefore divided into *true* and *false*.

TRUE HERMAPHRODITISM.—True sexual duplicity (*H. verus*) implies the existence of both the testicle and the ovary in the same subject, the condition of the external generative organs being of far less moment. In view of the manner in which the sexual glands arise, it is assumable that the indifferent anlage of the glands may undergo subdivision, one portion developing into the male, the other into the female organ, or the same may be accepted for the glands of the opposite sides. Theoretically true sexual duplicity may exist in three forms, bilateral, unilateral, and lateral (Klebs).

A. Bilateral hermaphroditism is the condition in which on both sides a testicle and an ovary exist, separate or partly fused into a composite organ. Almost the only instance of this malformation in man worthy of consideration has been the well-known case described by Heppner. In a two-months' child the external organs were of the masculine type, the penis being imperforate. The internal organs included a uterus, with tubes and ovaries on either side; likewise on each side an apparent testicle. While microscopical examination demonstrated the nature of the ovaries, it failed definitely to establish that the neighboring organs were testicles.

Very recently Garré[121] has reported a case which seems to merit the distinction of being regarded as a trustworthy bilateral true hermaphrodite. The individual, who sought the surgical clinic at Königsberg in Prussia, in addition to malformations of the external sexual organs, presented a hernia-like swelling in the left inguinal region. On opening this, various bodies and cords were found which proved to be testicle, epididymis, vas deferens, ovary, oviduct, and parovarium. The diagnosis was positively confirmed by microscopical investigation, the ovary containing numerous well-formed primary follicles and the testicle characteristic, although somewhat atrophic tissue, but without evidences of spermatogenesis. Digital examination through the rectum discovered on the left side two movable bodies, about the size of a pigeon's egg, together with a cord leading laterally to the urethra. These structures were assumed to be the left ovary and testicle with its duct.

B. Unilateral hermaphroditism is the condition in which on one side is a single sexual gland, on the other two—one testicle and one ovary. Two authentic cases presenting these peculiarities have been observed in the human subject, accepting the one described by Gast[122] as such. The latter was a still-born infant with exstrophy of the bladder. A rudimentary but well-developed penis was perforated by a urethra and lay between cutaneous folds. The internal organs were of the female type. The uterus was bifid and on the right was solid with a sheath-like fibrous extension, from which ran an oviduct with abdominal orifice. The left uterine division was also solid and bore an oviduct with fimbriated extremity. Attached to this was an ovary, showing follicles on microscopical examination; on the same side was a testicle, the size of a pea, from which a gubernaculum extended to the base of the left scrotal sac. Microscopical examination showed this organ to correspond to a testicle in its structure. On the right side no sexual gland could be discovered. The most convincing case, reported by Blanker and Lawrence,[123] occurred in a man. Microscopical examination proved the existence of an ovary and testicle upon one side, with the male gland upon the other.

C. Lateral hermaphroditism, in which one gland, testicle or ovary, exists on each side, is the most frequent variety of true double sex and the group within which the majority of reputed cases of true hermaphroditic malformations fall. Very few such cases, however, are substantiated by trustworthy microscopical examinations, and, therefore, are not beyond challenge. Among the most accurately investigated and convincing cases are

those reported by Rudolph,[124] Cramer, Meyer, Klebs,[125] Schmorl,[126] Obolonsky,[127] and Zimmermann.[128]

The case reported by Schmorl was that of an art student, aged twenty-two years, who applied to the surgical clinic in Leipsic for operation to correct hypospadias. The scrotum was rudimentary; on the right side was a small testicle, on the left none. The penis was small and drawn bow-like downward, with uncovered, well-formed but imperforate glans. The groove on the under surface of the penis was 3.2 cm. long, running into a small slit 0.5 cm. in length. An operation to correct the hypospadias and to free the penis was performed, but the patient shortly afterward died. The findings at the autopsy were as follows: Face bearded with hairs about 2 cm. long. Breasts undeveloped, but pubes had hairy growth resembling female. The penis, freed from its adhesions by the operation, measured 5.5 cm. in length with a circumference of 8 cm. To the sides of the penis were genital folds which projected above and grasped the penis between them. Opening into the urethra, 3.5 cm. behind the external orifice, was an aperture into which a probe could be passed for 15 cm. Further examination showed this canal to be a vagina and uterus, the latter consisting of a cervix and body. On the right side were a round ligament, tube and ligament analogous to that of the ovary, all of which extended down to the sexual gland in the right scrotal sac. This gland was shown by microscopical examination to be a testicle, although neither spermatozoa nor vas deferens were found. On the left side a tube ran into the inguinal gland and was continuous with an ovoid body, 5 cm. long and 2 cm. thick, which proved to be chiefly a distended and distorted fimbriated extremity. Histological examination revealed within this body a sexual gland having the characteristics of an ovary, but without ova.

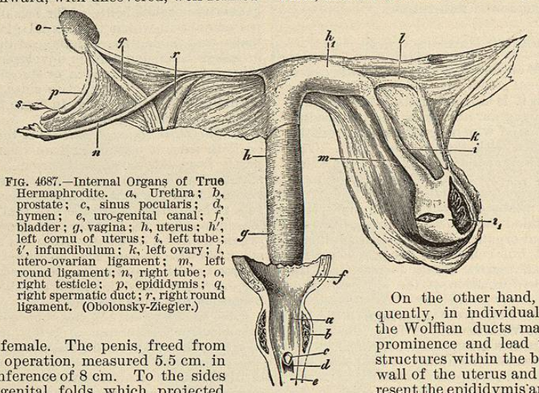

FIG. 4687.—Internal Organs of True Hermaphrodite. *a*, Urethra; *b*, prostate; *c*, sinus pocularis; *d*, hymen; *e*, uro-genital canal; *f*, bladder; *g*, vagina; *h*, uterus; *h'*, left cornu of uterus; *i*, left tube; *i'*, infundibulum; *k*, left ovary; *l*, utero-ovarian ligament; *m*, left round ligament; *n*, right tube; *o*, right testicle; *p*, epididymis; *q*, right spermatic duct; *r*, right round ligament. (Obolonsky-Ziegler.)

The case of Obolonsky was that of a twelve-year-old individual with external organs resembling a female with exceptionally developed clitoris. A well-developed vagina opened into the urethra at the sinus pocularis, guarded by a rudimentary hymen, and led to a unicornate uterus. A prostate was also present. The right broad ligament contained a testicle, an epididymis, and a vas deferens, as well as a rudimentary oviduct and round ligament. The left broad ligament contained an ovary with an ovarian ligament and a well-developed oviduct. Histological examination definitely established the nature of the sexual glands as testicle and ovary.

FALSE HERMAPHRODITISM.—As above pointed out, the crucial test in determining the existence of true double sex is the coexistence of both types of sexual glands; where the presence of these is not established by microscopical examination, the condition is to be regarded as one of pseudohermaphroditism, and the result of aberrant development of the excretory ducts and the external genital organs, which partially represent the characters of both sexes. The most frequent and striking of such malformations are those occurring in individuals of the male sex, in which, in addition to the usual structures, the Müllerian ducts, instead of being rudimentary and atrophic, have undergone unusually extensive development, resulting in the production of an enlarged uterus masculinus, or, in extreme cases, of a more or less well-formed vagina, uterus, and tubes.

In male subjects the external organs may be the seat of aberrant development, in consequence of which the parts assume female characteristics. Thus, the penis may remain rudimentary and resemble a clitoris. Following faulty union of the genital folds hypospadias results, and when extensive the urogenital sinus may open by an orifice resembling a vulva bounded by apparent labia—the ununited scrotal folds, the similarity being sometimes heightened by the absence of testicles in consequence of imperfect descent. Such malformations need not necessarily imply an hermaphroditic condition, although often a part of the defects observed in that connection.

On the other hand, but much less frequently, in individuals of the female sex the Wolffian ducts may assume an undue prominence and lead to the formation of structures within the broad ligament or the wall of the uterus and vagina, which represent the epididymis and vas deferens. Approximation and more or less extensive union of the genital folds, in conjunction with an abnormally large clitoris, may result in closure and obliteration of the vulva to such extent that the urogenital sinus opens beneath the apparent penis by a small aperture resembling a urethra.

Depending upon the predominating sex of the individual, as determined by the character of the sexual gland, pseudohermaphroditism is divided into the *male* and *female* type, in each of which group three varieties are recognized.

Masculine False Hermaphroditism.—I. *Internal*—in which associated with the normal, or nearly so, external male organs are a rudimentary vagina, uterus and, perhaps, tubes. The vagina pierces the prostate and opens into the urethra at the usual position of the sinus pocularis, or uterus masculinus.

II. *External*—in which the outer male organs exhibit faulty development resulting in imperfect fusion of the genital folds and ridges and the consequent resemblance to the female type. The disguise is favored by the general physical characteristics which are often distinctly feminine.

III. *Complete*—in which both internally and externally organs resembling those of the female are present. A more or less well-developed vagina and uterus, possibly also tubes, exist; the vagina opening into a rudimentary urogenital sinus, or short vestibule, into which may lead a groove from the under side of the penis. Less frequently the urethra and vagina open into the urogenital sinus by separate orifices. Very rarely these may be prolonged as canals in an otherwise normal penis, so that this organ possesses apparently double urethræ, the upper tube being the urinary channel, the lower one receiving the sexual ducts.

Of the large number of cases of masculine spurious sex duplicity in the literature the following case, reported by Stroebe,[129] is an interesting example of *pseudohermaphrodismus masculinus internus:* Autopsy on a male subject of sixty-three years showed normal external organs with the exception that the testes were undescended. Within the pelvis an elongated uterus, with well-developed body and fundus (6 cm. broad) lay behind the bladder. On the right side were found an oviduct,

a round ligament, and a sexual gland embedded within a broad ligament; on the left the tube was represented by a rudimentary cystic structure that probably was the remains of the Müllerian duct. A sexual gland was also discovered on this side. Microscopical examination showed the sexual glands to be atrophied testicles. The prostate was poorly developed. The uterus opened into the urethra at the sinus pocularis. The course of the vasa deferentia was interesting, since these canals in their lower part lay embedded on either side within the wall of the uterus. The seminal vesicles were abnormal, but the ejaculatory ducts were well developed. These findings point to the simultaneous development of the Wolffian and Müllerian ducts, leading to the production of excretory tubes of both sexes. Failure of the Müllerian ducts to undergo the usual regressive changes was manifestly the immediate cause of the abnormality; the reason for such failure, however, is less evident, although, as pointed out by Stroebe, the disturbing influences must be active at an early period.

Feminine False Hermaphroditism.—This condition is much rarer than in the male. Theoretically it may be subdivided into the three varieties—internal, external, and complete—described in connection with the masculine type.

I. *Internal*, in which, associated with normally formed external organs, the inner are augmented by derivatives of the persistent Wolffian ducts, the homologues of the epididymis and vas deferens, which lie within the broad ligament and the utero-vaginal wall.

II. *External*, in which the external organs assume the appearance of those of the male in consequence of more or less pronounced fusion of the genital folds. The modified parts often resemble a hypospadiac penis with cleft scrotum, the likeness to the latter organ being more striking when, as sometimes happens, the ovary descends through the canal of Nuck into the labium majus. In exceptional instances the urethra may be closed as far forward as the glans clitoridis, in which case the vestibule is no longer open and the external organs resemble those of the male.

III. *Complete*, in which both the internal and external organs present male characteristics. This condition is comparatively very rare. A well-developed prostate and even the homologues of the ejaculatory duct and seminal vesicle have been observed associated with persistent derivatives of the Wolffian ducts.

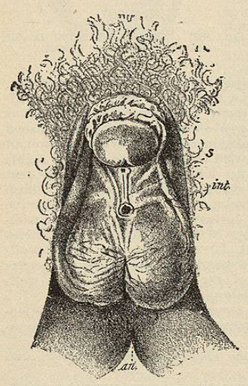

FIG. 4688.—External Organs of Male Pseudohermaphrodite. *s*, Groove; *int*, entrance of uro-genital sinus; *an*, anus. (Günther.)

The determination of the true sex in spurious hermaphroditism by observation upon the living subject is frequently uncertain, especially in view of the possible close resemblances of the external organs to the type opposite the real sex of the individual, as determined by histological examination of the sexual glands. In doubtful cases it is wise to assume that the patient is of the male sex. It should be remembered that spurious menstruation has been observed in masculine false hermaphroditism.

As an example of feminine pseudohermaphroditism established clinically, the case reported by Fitch[130] is of interest. A house servant, aged twenty-eight years, was subjected to examination, when apparently both male and female organs were discovered. The labia majora were of normal size; the labia minora were absent. In place

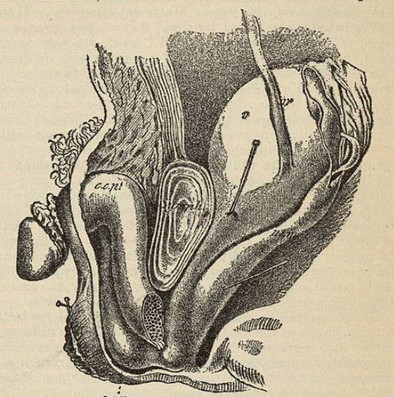

FIG. 4689.—Internal Organs of the Preceding Case. *Symph.*, Symphysis; *v*, bladder; *utm*, uterus masculinus, greatly enlarged; *ur*, ureter; *ccp*, corpus cavernosum; *ccu*, corpus spongiosum. (Günther.)

of the clitoris of usual size was an apparent penis, which, when in erection, measured five and a quarter inches in length and three and three-eighths inches in circumference. The glans was perfect and provided with a urethra. The scrotum was about two inches long and seemingly contained two testicles (?). It was claimed that semen (?) was ejaculated from the urethra. The vagina was spacious and had an os uteri projecting into it. Seven years before the subject had given birth to a normal female infant. Scanty menstruation occurred every three weeks and lasted two days. Sexual gratification was said to be equally distributed between the two sets of organs.

Marchand[131] has described a case of feminine pseudohermaphroditism of exceptional interest as illustrating possible sources of error unless careful histological examination is made of supposed sexual glands. The clitoris was large and but slightly hypospadiac. The vagina opened into the urethra, which was surrounded by a small prostate. Both oviducts were closed at their distal end. Very small ovaries were present in their normal position; that these organs were of this nature was determined by microscopical examination. On the right side the outer border of the broad ligament contained a second body of the size of a well-developed testicle, which proved, however, to be not a sexual gland, but an enlarged accessory suprarenal. Since the presence of these bodies, of very small size, is to be regarded as constant, it is probable that when enlarged they have been mistaken for sexual glands, hence their possible hypertrophy must be borne in mind. Those interested in the literature of the various types of hermaphroditism will find in Taruffi's monograph[132] an exhaustive and systematic review, arranged chronologically.

MALFORMATIONS OF THE EXTREMITIES.—Congenital defects involving the limbs include two chief groups: (1) Those following arrested development, and (2) those occurring in consequence of excessive development, the former resulting in deficiencies, the latter in redundant parts of limbs, especially the digits.

The first appearance of the limb-buds in man occurs during the third week of embryonic life, as flattened rounded elevations which project laterally from the body

wall. During the fifth week these growing processes exhibit differentiation into proximal and distal segments, the latter becoming the hands and the feet. By the close of this week the distal segment shows beginning division into digital areas, the upper limbs anticipating in their differentiation the lower by some days. The proximal segment soon becomes subdivided, so that by the sixth week the three chief segments of each limb are evident. The seventh week brings greater distinctness in the separation of the digits and indication of the carpal region of the hand and the corresponding parts of the foot. With the eighth week appear the elbow- and knee-joints. The digits, although advanced in development, are still webbed, being connected by a membranous fold of integument. By the ninth week the several divisions of the limbs are present, but not in their definitive proportions. Thus the length of the entire hand is excessive in relation to that of the arm and forearm; likewise, the fingers are too long in proportion to the remaining parts of the hand, and the thumb too long for the other fingers. As already noted, the upper extremity at first leads the lower in its differentiation, but by the ninth week both are of equal length. From this time, however, the lower limb begins to outstrip the arm in its length growth.

The primary shovel-like form of the limb-bud projects at right angle to the body wall; later, with the differentiation into segments, the limbs become folded ventrally and parallel to the sides, stretching obliquely from head tailward. At this stage the future flexor surface of the limb is presented ventrally, the extensor surface dorsally. The preaxial borders of the limbs (radial and tibial, marked by the thumb and great toe) are directed toward the head; conversely the postaxial borders (ulnar and fibular) look tailward. During subsequent growth the upper and lower limbs undergo partial rotation, the arm turning outward and backward, the leg inward and forward. In consequence of these opposite rotations the preaxial border of the upper extremity (thumb and radius) is carried laterally and dorsally, while the corresponding border of the lower limb (great toe and tibia) is turned medianly and anteriorly, these parts of the two extremities being homologous notwithstanding their apparent dissimilarity in the completely developed condition. In their differentiation from the mesodermic tissue composing the early limb, the skeletal segments follow the order of their relations to the trunk, the proximal segments being first defined, the distal ones last.

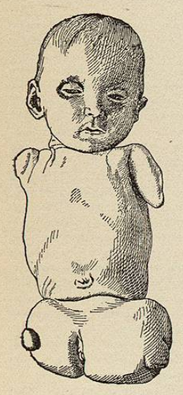

FIG. 4690.—Amelus. (Ziegler).

Congenital Deficiencies of the Extremities.—These malformations are attributable to causes of several kinds, some of which lead to primary defects, others to secondary. Among the first are primary deficiency in the primitive limb anlage, perhaps inadequate differentiation of the segments and deficient bone formation. The most important causes producing secondary malformations — those modifying limbs which under favorable conditions would develop into normal members— are inadequate intra-uterine space, thereby inducing pressure, and amniotic adhesions and constrictions. Defective development of the extremities is sometimes associated with profound congenital malformations of the nervous system, as spina bifida, involving the nerve trunks supplying the limbs.

The classification of malformations of the extremities suggested by Kümmel,[133] also followed by Kalussner,[134] is the most consistent grouping based upon anatomical data, and to those especially interested in these defects the monographs by the above authors are of particular value. For the purposes of the general reader, however, the clinical classification is more convenient and, while less exact, presents the defects in a useful sequence; it has, therefore, been followed in these pages.

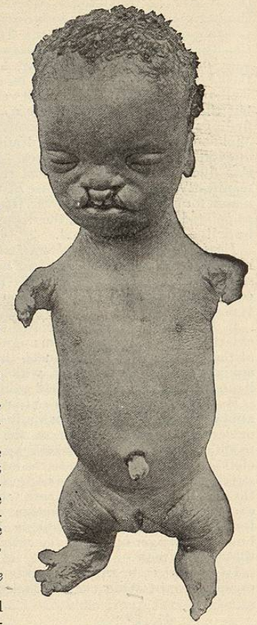

FIG. 4691. — Phocomelus with Double Harelip. (Hirst and Piersol.)

I. *One or more limbs entirely wanting.*

1. *Amelus.* Both upper and lower extremities are absent, while the trunk is usually well developed. The position of the limbs is often indicated by button-like elevations within a funnel-shaped depression.

2. *Abrachius and Apus.* This condition implies the suppression of either the upper or lower limbs, the remaining pair being often well developed.

3. *Monobrachius* and *monopus* result from absence of a single arm or leg, the other limbs, including the mate to the missing member, may be normal.

II. *One or more limbs defective.*

1. *Peromelus.* All the limbs are imperfect, although the causes have not been so radical in their action as to result in complete absence of the extremities. The degree to which the upper and lower limbs are affected often varies, thereby producing the conspicuous malformation known as *phocomelus.* This is distinguished by the rudimentary condition, or complete suppression, of the proximal segment with relatively well-formed hands and feet. According to Kümmel, the most frequent combinations are (*a*) defective development of thigh with relatively well-developed leg and foot; (*b*) defective development of arm or forearm with comparatively well-formed hand. A defect of the upper extremity corresponding to (*a*), that is, defective arm with well-developed forearm and hand, probably does not occur; likewise suppression of the thigh and leg with well-formed foot is very rarely observed.

2. *Perobrachius* and *peropus* are represented by defective development affecting the upper and lower extremity respectively.

III. *One or more limbs abnormally small, although well formed.*

1. *Micromelus,* in which all of the extremities are deficient in size, the diminution, however, being unattended by malformation.

2. *Microbrachius* and *micropus* include such cases in which the abnormality is limited to one or both arms or legs respectively.

IV. *Limb-bones defective or absent.*

Congenital imperfection or absence of the bones of the arm or thigh, forearm or leg, produces a series of ray defects in which malposition and faulty development

of the succeeding segments are usually conspicuous, this being particularly so in the hands and feet in conjunction with the absence of the radius and tibia. Defective fibulæ are generally associated with more or less marked shortening of the limb.

V. *Lower limbs fused.*

Sympus, symmelus, or *siren,* as such malformations are variously termed, is distinguished by more or less complete fusion of the lower extremities. The lower end of the trunk is also defective, as evidenced by the malformations of the pelvis and of the external genital organs and excretory passages which are commonly imperfect. The united limbs usually have undergone rotation outward and backward, so that the primary external surfaces become fused. In consequence of such union the lower half of the body forms a conical mass, which may terminate below in a rudimentary single foot (*Sympus monopus*), or two imperfect feet may be present (*S. dipus*). At other times only one or more stunted toes are seen, or all traces of feet are wanting (*S. apus*), the fused extremities ending in a rounded, somewhat elongated apex.

VI. *Hands or feet defective.*

Malformations of the distal limb segments due to defective or arrested development, or *perodactylism,* occur in great variety as to form and detail. In general these abnormalities result from the deficient formation, or entire suppression, of one or more phalanges, combined frequently with fusion of certain digits (*syndactylism*). The variations in these defects are so great that they include all degrees of imperfection, from the scarcely perceptible shortening of a single finger to the presence of mere stunted knob-like digits. One form of malformation, *cleft hand or foot,* is conspicuous on account of the forked appearance due to separation of the marginal digits by a cleft resulting from the imperfect development, or entire absence, of the intervening metacarpal bone or bones and the directly related carpals. The phalanges of the third finger, with the associated metacarpal and carpals, or the corresponding bones of the foot are most frequently wanting, the remaining digits on either side being fused into a marginal ray. In some cases heredity is apparently an important factor in the production of these malformations, as shown in the striking instance of hereditary brachydactyly recorded by Webb,[135] six generations of a family being "short-fingered."

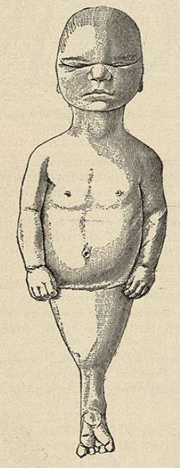

FIG. 4692.—Sympus Dipus. (Ziegler.)

Syndactylism, the congenital union of two or more digits, may be primary or secondary. In the former case the fingers or toes are joined in consequence of imperfect primary differentiation and separation, the digits remaining connected by robust intervening bridges, at times appearing merely as ridges projecting from the fin-like terminal segment. Such malformations are manifestly the result of developmental arrest in consequence either of defective primary formation or of unfavorable influences subjecting the developing digits to unusual pressure. In extreme cases, when all the fingers or toes are blended, the union is usually very intimate, the hand or foot constituting a terminal knob-like mass presenting little resemblance to the normal form. Secondary syndactylism results from more or less extensive union of the digits subsequent to their development. In such cases the connection usually consists of integumentary folds extending for a variable distance from the bases of the digits toward their tips. Exceptionally the union is limited to the latter, the bases remaining free, thereby producing the fenestrated variety of syndactylism.

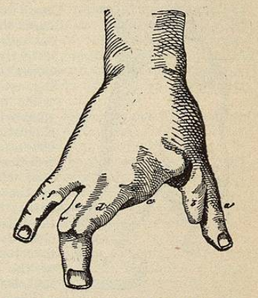

FIG. 4693.—Malformed Hand (Perochirus) with Fusion of Fingers. (Otto.)

Spontaneous or *congenital amputation* of the extremities, effected by constricting cords or bands of amniotic tissue, is responsible for numerous defective limbs. When the ligatures have been insufficient to produce necrosis and complete division of a member, their position is often indicated by marked constrictions and their baneful influence is manifested by the stunted but still attached distal portion of the limb. The latter being compressed within the encircling folds of amnion is unable normally to develop, remains in the fœtal condition, and fails to keep pace with the growth of the more favored parts of the extremity. Spontaneous amputations may involve any part of the extremity, from a part of a single digit to loss of an entire limb. The latter usually ends in a smooth conical stump, while the amputated portion, when separation has occurred at an early period, is commonly not to be found, having undergone complete maceration and disappeared.

Congenital Luxations and Malpositions.—Although the anatomical peculiarities exhibited by osseous and ligamentous structures involved in congenital luxations are usually secondary results and not primary causes of these malformations, the latter call for brief notice in connection with defects of the extremities.

Congenital luxations of the femur are the most important as well as frequent of the prenatal dislocations. They are always secondary and probably due to the effects of malposition of the limbs at a period before the joint was capable of resisting the unusual strain, in consequence of which the femur is forced from its normal socket. Loss of apposition and function result in atrophy and malformation of the femoral head and the acetabulum, the latter being often small and shallow and at times almost wanting. The capsule is stretched and abnormally large. The luxation may be on one or both sides. Similar deformities may affect the knee-, shoulder-, or elbow-joint, although much less frequently.

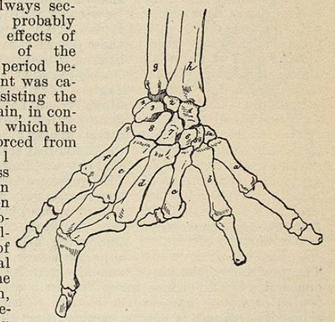

FIG. 4694.—Skeleton of Preceding Deformed Hand. (Otto.)