two instances recorded by Preuss and by Ahlfeld. In the infant observed by the latter, the motion was produced by contractions of a sheet of striped muscle beneath the covering of the tumor and not by true fœtal movements.

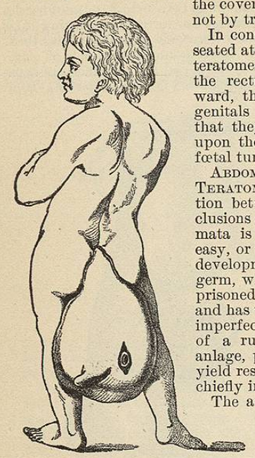

FIG. 4673.—Child with Sacral Tumor. (Löffler.)

In consequence of the deep-seated attachment of the sacral teratomes, the perineum and the rectum are pushed forward, the anus and extended genitals being so displaced that they usually lie directly upon the anterior part of the fœtal tumor.

ABDOMINAL PARASITES AND TERATOMATA. — The distinction between true parasitic inclusions and embryonal teratomata is by no means always easy, or indeed possible. The development of an independent germ, which has become imprisoned at an early period and has undergone only a very imperfect evolution, and that of a rudimentary embryonal anlage, producing a teratoma, yield results which often differ chiefly in degree.

The anlage of the parasitic inclusion may lie, theoretically at least, embedded within the abdominal wall of the autosite, or it may be included within the body cavity. The entire number of adequately described and undoubted abdominal fœtal inclusions is small, probably less than forty. As an example of the very rare intraparietal type of inclusion the case mentioned by Himly [74] stands almost alone. Autopsy of a female child of two and one-half years revealed a cystic tumor in the epigastrium which contained a fairly well-formed fœtus, with, however, defective extremities and head.

The intra-abdominal inclusions, or *engastric parasites*, are found most commonly in the vicinity of the lesser peritoneal sac, between the layers, or at the root, of the transverse mesocolon, being an especially favorite location. Since, as shown by Paquy, [75] the true fœtal inclusions always lie supra-umbilical and closely connected with the alimentary canal, Lexer [76] regards their usual intimate relation to the lesser peritoneal sac as due to migration induced by the increasing volume of the liver and the transverse displacement of the stomach. The fœtal parasite is usually enclosed within an envelope, composed of mesenteric or retroperitoneal tissue, to which the rudimentary embryo may be attached. Sometimes the fœtal characters are so well pronounced that spine, head, and extremities are recognizable; in other cases only rudimentary organs are present. The sac surrounding the inclusion is closely united with the adjacent organs, the blood-vessels of which communicate with those of the parasite and provide for its nutrition. Not infrequently the rudimentary fœtus exhibits distortion, the result of progressive mechanical disturbances as well as of faulty development. Under such conditions the inclusion may become pressed into a shapeless mass.

Spontaneous movements have been noted within the cyst containing the parasite. Klebs [77] describes a case of an infant in which independent motions were noted within an abdominal tumor. The child died a few weeks after birth, and presented, on post-mortem examination, a cyst composed of peritoneum and amnion beneath the transverse colon. An artery passed from the aorta of the host to the cyst and thence by a short umbilical cord to the parasite. The latter was of an irregular ellipsoidal form and consisted of an imperfect spine and pelvis, a skull of considerable size containing a distinct brain mass, a digestive tube with unusually wide, large intestine, a liver, and rudimentary lungs and heart. The extremities were represented by poorly developed and stunted limbs. External genitals were absent.

Another remarkable instance of abdominal fœtal inclusion is the classic case recorded by Highmore. [78] The subject, when a boy of seven, developed a marked abdominal swelling attended with severe pain. Succeeding a period of temporary relief until the fifteenth year, the distress returned, death occurring in consequence of intestinal hemorrhage. During life marked pulsations were recognizable in the tumor, sensitive on pressure; the movements were felt by the patient, who declared that his abdomen contained something living. The autopsy disclosed an irregularly oval, sack-like tumor, weighing over four pounds, attached along the entire extent of the duodenum, which freely communicated with and formed a part of the sack. The latter contained an imperfect fœtus with stunted trunk, two upper and one lower extremities, and a very rudimentary head consisting of scalp to which hair a foot long was attached. The nutrition of the parasitic fœtus was maintained by a short cord which established the connection with the vascular walls of the sack.

Ballantyne [79] describes an abdominal teratoma in a child of three months, the parasite being connected with the lesser peritoneal sac. At one point there was a projection resembling a pair of rudimentary digits; pieces of intestine, teeth, and fragments of bone and cartilage were also present. Paquy records a somewhat similar case in which the omentum was occupied by a tumor consisting largely of cartilage, glandular elements, and abundant blood-vessels.

Although children bearing fœtal inclusions are often still-born, the possession of the parasite is not incompatible with prolonged, and even extended, life, since such inclusions have been discovered in subjects of advanced years. Usually, however, the presence of the parasite induces, after a time, fatal peritonitis.

EMBRYONAL TUMORS OF THE SEXUAL GLANDS.—The sexual glands are the seats of fœtal tumors of great interest by reason not only of their relative frequency, but also of the interesting problems concerning their production. Disregarding for the present the latter, these tumors may be divided into *cystic dermoids* and *teratomata* according to their consistence—sac-like or solid respectively—and the degree of development which they present.

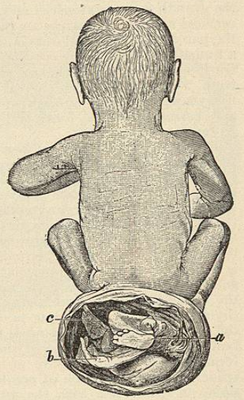

FIG. 4674.—Sacral Teratoma Containing Extremities (*a*, *b*, *c*) of Parasite. (Ziegler.)

Ovarian cystic dermoids, the more common variety of the first group, appear as tumors varying in size from a pea to a child's head. These consist of a tough-walled sac enclosing a fatty mass and hair. The seat of the dermoid is nearly always the sexual gland, the tumor being attached to the adnexa of the uterus by broad adhesions. The ovarian tissue immediately surrounding the growth usually undergoes partial or complete degeneration, although in exceptional cases the dermoid may be almost isolated and connected with the ovary by only a slender stalk. Wilms [80] has established the important

fact, confirmed by a number of competent observers, that the embryonal tumors of the sexual glands contain derivatives of all three germ layers, and further, that the development proceeds in general, although modified, in a way similar to that followed under normal conditions.

The inner surface of the dermoid cyst at some point exhibits a projection, villous or less abrupt, which usually bears hairs and also not infrequently teeth. While the more superficial layer of the projection contains the constituents of the integument, including hair follicles and sebaceous and sweat glands, the deeper parts may enclose cartilage, bone, muscular tissue, nervous substance, mucous glands, or suggestions of the digestive and respiratory tubes. Rudimentary mammary glands with nipples, fingers with nails, and retinal pigment are among the rarer additional organs that have been observed in ovarian dermoids. Hepatic, renal, and cardiac tissue, on the contrary, are among the few structures not represented in these tumors. The explanation of these exceptions, as suggested by Wilms, lies in the early assumption by the autosite of the nutrition of the parasitic embryo, which results, after a possible abortive effort at heart formation, in a very imperfect independent circulatory apparatus. In consequence of this relation there is no necessity for the formation of excretory organs, hence the liver and kidneys remain undeveloped and are never found.

Although most frequently encountered in middle life, ovarian cystic dermoids occur at all ages from early childhood to extreme old age.

Ovarian teratomata, or solid tumors, are of much greater rarity than the cystic dermoids. As the latter, so also the teratomes always contain the representatives of all the germ layers. The tissues and rudimentary organs, however, present much less regularity in order and arrangement and hence less resemblance to the fœtus on account of their confused and indefinite disposition and generally less perfect development. These characteristics are due to the mechanical disturbances induced by limitations of space, in consequence of which there follows a displacement of the cells resulting in loss of relations in the anlage and the institution of a process of tumor formation. As shown by Wilms, [81] the solid ovarian teratomata correspond in genesis with the cystic dermoids and differ only in the degree to which the fœtal structures are developed. In view of the slight resemblance that these growths bear to the normal products of conception, Wilms has appropriately named them *embryonoid tumors*.

The *fœtal growths of the testicle* constitute a group homologous with those of the ovary already discussed, in conjunction with the latter forming the distinct class of rudimentary parasites of the sexual glands. Wilms conclusively proved that these growths originate from the testicle and correspond in all important features with those of the ovary. As in the latter so also in the testicle dermoid cysts and solid teratomes both occur; in the male gland, however, in contrast to the female, the cystic dermoids are of extreme rarity, the solid tumors being the form usually encountered. Even these are of great infrequency when compared with the ovarian teratomata. As pointed out by Wilms, [82] in the testicle the conditions favorable for the development and growth of fœtal structures are seldom present on account of the anatomical construction of the organ and its position, hence the formation of rudimentary parasites but rarely takes place, the solid embryonoid tumors being the usual expression of the repressed development.

Dermoid cysts of the testicle, although of great infrequency, occasionally present marked fœtal characters, as in the remarkable case reported by Répin [83] in which an almost complete skeleton, with four limbs, was present. The more usual teratomes evince their embryonal nature by the presence of tissues derived from the three germ layers rather than by an orderly disposition suggesting parts of a fœtus.

These tumors vary greatly in size, the largest being in volume equal to a child's head. They may be entirely solid or contain cysts which may be lined by simple or stratified cuboidal or cylindrical epithelium; in rare cases pigmented as well as glandular cells have been found. The derivatives of the mesoblast are conspicuous in these tumors, being represented often by considerable masses of connective tissue and cartilage, less frequently by bone; adipose and muscular tissues are also not uncommon. The teratomata of the testicle, therefore, are usually met with in the form of adenocystoma, chondroadenoma, chondrosarcoma, adenorhabdomyoma, or other type of composite tumor.

The studies of Wilms leaves no doubt that these teratomata are to be regarded as imperfect expressions of an independent development, and that they in principle are to be regarded as extremely rudimentary parasitic malformations modified by the influence of mechanical disturbance.

ORIGIN OF PARASITIC DUPLICITIES.

The condition of true parasitic duplicity, as distinguished from dependence secondarily acquired by one of the united twins in consequence of unfavorable conditions of development, implies *primary inequality* of the anlages.

In all forms of double monsters the allantoic circulation of one individual may become impaired or interrupted to such extent that the affected embryo succumbs unless its nutrition is assumed by the more fortunate mate. The donation by the latter may so far suffice that a considerable part of the dependent embryo is maintained, thereby producing such striking forms as "Laloo." When, on the other hand, the union is cranial and the donated nutrition insufficient, only the cephalic end may remain, as in the classic case of Home, in which a well-developed head, attached to the vertex of the autosite, alone represented what was at one time a complete embryo.

The secondary and acquired nature of such parasites is conclusively demonstrated by the presence of the remains of an atrophic umbilical cord attached to the trunk. Since the nutrition of the affected embryo is early assumed by the stronger, the heart of the former atrophies, although instances have been noted in which the hearts of both parasite and autosite have existed within the thorax of the autosite.

The true parasitic duplicities are from the first conspicuously asymmetrical, the dependent embryo being rudimentary or incompletely developed, and more or less enclosed by the body of the autosite. These inclusions present two groups: (*a*) those which are partially surround and hence appear as an external appendage to the host, or are drawn within the body and lie beneath the integument of the autosite; (*b*) those which develop entirely within the autosite, especially in relation to the abdominal cavity. Examples of the first group are seen in the epignathi and the sacral tumors; of the second in the ovarian dermoids.

The exact manner in which the weaker and rudimentary anlage becomes surrounded by the stronger can only be surmised, but it may be assumed with probability as intimately related with the migration of the external layer of the autosite which takes place in connection with the invaginations and the closures incidental to the normal development of the stronger embryo. Such closures occur at the cephalic pole and along the dorsal and ventral surfaces, while the primary oval and branchial invaginations are additional means of transportation. We may reasonably conceive the anlage giving rise to the future parasite as being fixed to the surface and within the amnion of the autosite in the immediate vicinity of such migratory areas; subsequently it is carried within the body and more or less surrounded by the tissues of the autosite to produce a true fœtal inclusion, or *fœtus in fœtu*.

The production and relations of an epignathus or encranius can be clearly referred to the inclusion of anlages which have suffered displacement during the formation

of the primary oval invagination and of the brain case respectively. In other cases, however, the location of the fœtal inclusion is probably acquired secondarily in consequence of changes occurring during development, as in the case of sacral teratomata on the ventral aspect of the sacrococcygeal region in which the anlage was

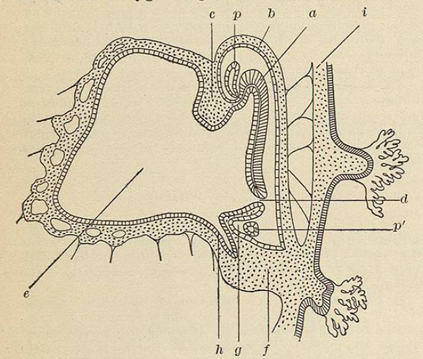

Fig. 4675.—Schematic Representation of Relations of Parasitic Anlage in Epignathus (*p*) and Sacral Teratoma (*p'*), based on sagittal section of Spee's early human embryo. *a*, Neural plate; *b*, amnion; *c*, heart area; *d*, neurenteric canal; *e*, umbilical vesicle; *f*, belly-stalk; *g*, region of anal membrane; *h*, allantoic duct; *i*, chorion. (After Marchand.)

primarily attached in the vicinity of the hind-gut and carried ventrally by the flexure of the autositic axis.

In contrast to the primary independence of the nutritive apparatus of both embryos in the case of parasites arising from symmetrical double monsters, the rudimentary anlages producing true fœtal parasites present little evidence of having had separate chorionic attachments. It is doubtful whether such parasites ever possess rudiments of true umbilical cords or allantoic attachments, notwithstanding their reported presence in certain cases.

In attempting to arrive at some conclusion regarding the vexed question concerning the origin of the rudimentary anlage producing the parasite, it may be assumed that the latter represents the development of a germ-cell or cell mass that progresses independently in the earliest stages, although later dependent for its nutrition upon the autosite. Reviewing the established facts relating to the origin and early changes of the ovum leading to the formation of the blastoderm from which the embryo proceeds, two possible sources of the rudimentary anlage may be suggested that are not incompatible with the known facts of embryology. These are: (1) that the rudimentary anlage may be the product of the imperfect development of a fertilized polar body; (2) that the parasite may arise from the differentiation of a special or an isolated group of segmentation cells.

The significance of the polar bodies as abortive or rudimentary ova, liberated during the process of maturation of the female sexual cell, is admitted by all. These rudimentary ova, the polar bodies, contain the same morphological constituents and therefore the same potentialities as the larger matured oöcyte, only in diminished force. Usually the polar bodies remain for a time beneath the egg envelope, or zona pellucida, closely related to the early segmentation spheres, later disappearing.

Since the occurrence of segmentation and formation of the blastoderm in vertebrates implies the development of a fertilized ovum—a true parthenogenetic development not occurring among the vertebrated animals—it is of interest to note that in addition to the direct observations of Platner and of Kostanecki on the fertilization of the polar bodies of lower forms, the older observations of Bischoff, proving that in the segmenting ovum of mammals (rabbit, dog) the living spermatozoa may exist beneath the zona pellucida in the vicinity of the polar bodies, have been confirmed by Assheton [84] and by Bonnet.[85] In view of these investigations it is highly probable, at least not improbable, that in exceptional cases the polar body may undergo fertilization and enter upon a development which may produce a small blastodermic vesicle and rudimentary anlage capable of giving rise to the parasite. The anlage so arising lies closely related to the blastoderm developed from larger egg. When that derived from the polar body retains the usual primary relations of the latter it will lie in proximity to the first plane of cleavage, which corresponds with the later axis of the embryo as established by the notochord and the medullary furrow. Since the last-named structure becomes the neural tube, it is evident that the rudimentary anlage may become attached in the vicinity of the medullary folds and be drawn within the larger embryo to become an encranius. It is, however, well established that the polar body may lie in relation to other portions of the segmenting mass and hence become attached to various parts of the larger anlage. When related to the ventrally approximating somatopleuric folds, the rudimentary anlage may be enclosed within the later abdominal walls or carried into the body cavity to form an inclusion.

The possibility of the rudimentary anlage being early included between the segmentation cells of the larger anlage has been shown by the observation of Assheton on the rabbit in which the polar body was found between the blastomeres. Assuming that the ferilized polar body suffers such displacement it follows that the resulting rudimentary anlage may obtain later an intramesoblastic position and may become an inclusion in some deeply seated organ, including the sexual glands. In such manner the existence of the ovarian dermoids and the teratomata of the testicle find a plausible explanation.

The important fact that the embryonoid growths of the sexual glands consist of derivates of all three germ layers was emphasized by Wilms in the papers already quoted. Other investigators, including Pfannenstiel, Krömer, Mertens, Arnsperger, Franzen, and Emanuel, have confirmed these views, which may be regarded as established. Bandler,[86] however, challenges these conclusions and denies that the entoblastic derivatives take part in the formation of these growths, the origin of which he attributes to possible cell dislocations occurring during the complex development of the Wolffian body, kidney, and sexual glands. The formation of such growths implies development of a sexual cell. The necessity of accounting for such process has led to many theories as to the possibilty of development proceeding from sexual cells within the ovary and testicle without the influence of the male element. The well-known existence of parthenogenic development among certain invertebrates suggested the possibility of the occurrence of a similar process in the higher animals; parthenogenesis, therefore, has been accepted as an explanation of the congenital tumors of the sexual glands by many authors, some going to such length that they accept the radical proposition that the sexual cells within the testicle are capable of producing the teratomes of that organ. Additional support for such views has been claimed from the interesting experiments with unimpregnated eggs of certain invertebrates which under particular conditions undergo cleavage and attempt the earlier phases of development. Somewhat analogous changes have been observed in the ova of vertebrates resulting in an imperfect segmentation, or more accurately "fragmentation." The conclusion of Bonnet,[87] that there is no justification for assuming the occurrence of parthenogenesis among the vertebrates, may be accepted as a reliable verdict founded upon an exhaustive and critical review of the evidence presented by the literature of this interesting problem.

Beard [88] has suggested an explanation of the congenital

tumors of the sexual glands, as well as of other regions, based upon his investigations on the early embryos of the skate. In these he found that particular blastomeres are very early set apart for the formation of the sexual cells of the embryo and take no part in its general development. The descendants of these special *primary germ-cells* constitute the sexual elements of the ovary and the testicle. Beard regards the embryonal tumors as derived from aberrant members of this group of primary germ-cells, which differ from the generations of secondary germ-cells, represented by the ova and the spermatozoa, in not having undergone the cytological changes incident to maturation, extrusion of the polar bodies, and reduction of chromosomes. The primitive germ-cell, as Beard regards it, is the sister, not the derivative, of the embryo, and therefore capable of giving rise to a rudimentary twin — the included parasite or embryoma. As Beard admits, he establishes only the existence of the vagrant primary germ-cells and does not explain their spontaneous development. He regards these cells, however, as endowed with an inherent potentiality which may institute independent formative processes, so that "neither a congregation of germ-cells nor a parthenogenetic development of such is needed to account for dermoids."

A possible source of the rudimentary anlage producing the parasite has been suggested by the interesting experiments relating to the development of separated segmentation spheres, carried out by various investigators during the last decade. The observations of Driesch, Born, Wilson, Morgan, O. Hertwig, O. Schultze, Herlitzka, and others, have shown that artificially separated segmentation spheres may develop into perfect although small embryos, the size, within certain limits, being determined by the proportionate value of the isolated cells to the entire segmentation mass. The observations of Roux [89] upon young frog larvæ seem to warrant the assumption that groups of segmentation cells may become disconnected and displaced and finally enclosed within the derivatives of the blastodermic layers. Bonnet assumes, and with reason, that such inclusions may later undergo further differentiation and become congenital fœtal growths. Coley and Buxton conclude, after reviewing the various theories, that the separation of segmentation cells offers the most satisfactory explanation.

There are, therefore, two plausible sources for the rudimentary anlage producing the parasite, the one being the fertilization of an abortive ovum (the polar body), the other the tardy differentiation of a group of vagrant segmentation cells. There seems no reason for regarding either of these possibilities as the exclusive mode of production, and it may be assumed with probability that both methods may contribute to the various types of parasitic duplicity. The observed attempts, in certain instances, at the formation of imperfect fœtal membranes may be related, possibly, with an origin from a fertilized polar body, the development of which abortive ovum would be more favorable to the formation of such structures than that of included segmentation spheres.

The foregoing considerations emphasize the common origin of cystic dermoids and teratomes of the sexual glands and of other fœtal inclusions, the sharp lines drawn by Wilms being justly regarded by Marchand and by Bonnet as unwarranted.

The causes determining the period at which the fœtal inclusion shall develop within the surrounding tissue remain unknown. In general it may be assumed that conditions affecting nutrition and tissue activity are responsible. The fact that the ovarian inclusions usually develop during the period of sexual activity may be attributed to the stimulus incidental to the well-known periodical congestion of the ovary and associated parts. On the contrary, the fact that teratomata of the testicle are more frequently encountered in young subjects may be connected with the more favorable conditions of active nutrition presented by the young organ than during the later years where the imprisoned anlage suffers compression and arrest.

Malformation of the Organs.

The malformations described in the foregoing pages are the expression of influences affecting more or less profoundly the entire organism; it remains briefly to discuss the more limited developmental defects that are encountered in the individual organs.

Defects of the Cerebro-Spinal Axis.—The early appearance, exposed position, and delicate walls of the neural tube render it especially liable to unfavorable influences affecting its growth which result in malformations of the spinal cord and brain and their membranous and bony envelopes.

It will be recalled that the cerebro-spinal nervous axis is the direct transformation of the neural tube, the latter first appearing as a groove along the dorsal surface of the early embryo. The ectodermal medullary folds bounding the intervening neural groove approach and gradually convert the furrow into the neural tube, the expanded cephalic segments of which form the brain vesicles. Following the closure of the neural tube by fusion of the medullary folds, the surface ectoderm becomes separated from that lining of the neural tube by the ingrowth of the surrounding mesoderm. The latter subsequently undergoes differentiation into the meninges and the vertebral column and brain case. Each segment of the spine, consisting of a body and a pair of neural arches, is developed within the mesoblast, supplied by the somites as the skleratome, from three chief centres. The latter are: one for the body and one on each side for the corresponding half of the arch. Failure of the arches to unite dorsally in the midline may result in a more or less conspicuous cleft associated with developmental disturbances of the spinal cord and its membranes.

Defects Caused by Imperfect Closure of the Spinal Canal.—Clefts of the vertebral canal may be divided according to their relations with the exterior into two chief groups, *open* and *covered*. The former, termed *rachischisis*, present an open furrow-like defect at the bottom of which may be seen the vertebral bodies. The covered clefts are distinguished by the presence of a more or less prominent sac, hence the names *spina bifida cystica*, or *rachischisis cystica*, by which this group is properly known, although the less specific term *spina bifida* is often used instead. Both forms of cleft may occur in any part of the spine, and may be limited or very extensive.

Rachischisis.—The deformity included under this name is due to faulty development and imperfect union of the neural arches. When well marked it appears as a widely open groove bounded by rudimentary processes on each side representing the imperfectly formed arches. The skin being absent, the vertebral bodies are covered by the dura and the ventral portion of the pia, with, perhaps, remains of the atrophic spinal cord and the associated nerves. The cleft may involve the entire canal (*holorachischisis*), and is then frequently associated with an analogous defect of the skull; or it may be limited to one part of the spine (*merorachischisis*). The usual seat of the latter defect is the lumbo-sacral region, although clefts in the cervical portion of the spine occasionally occur. The intervening thoracic region is seldom involved. Rachischisis is usually attended with spinal curvature; the total and cervical clefts are associated with lordosis and those of the lower segment with kyphosis. In the latter cases, other profound defects of the ventral body wall, as eventration or ectopia of the bladder, are often also present.

Viewed from behind, the centre of the defect is occupied by a soft red membrane; this represents the flatly expanded pia enclosing the remains of the rudimentary spinal cord which has succeeded the unclosed neural tube. When the rachischisis is total, the cord is practically absent (*amyelus*); when less extensive, the cord may be present as a plate-like strip of loose, highly vascular tissue fused with the pia. This mass, the *area medullo-vasculosa*, may form a continuous structure, or be broken up into a delicate veil-like tissue. Remains of nerve cells

and nerve fibres may be recognizable. At its margin the central area becomes continuous with the surrounding integument, the epidermis of the latter extending for a variable distance upon the pia. This boundary ring is

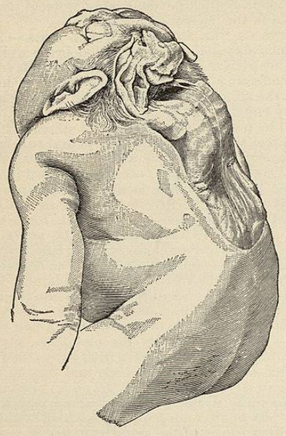

FIG. 4676.—Cranio-rachischisis with Absence of Brain and Spinal Cord. (Ziegler.)

termed the *zona epithelioserosa*, outside of which lies the *zona dermatica*, as the thicker margin of the surrounding integument is called.

The ventral relations of the meninges are more normally preserved, the anterior part of the subarachnoidal space being represented by the cleft between the pia and the subjacent arachnoid attached to the dura. This space sometimes becomes distended by accumulation of fluid. The rudimentary spinal nerves are attached to the ventral surface of the vascular membrane and may at times be followed into the atrophic nervous mass. In rare cases the vertebral bodies themselves may be imperfect and cleft, thereby giving rise to *rachischisis anterior*.

Spina Bifida Cystica.—The covered forms of spinal cleft are distinguished by the presence of a cyst, which protrudes through the fissure in the spine, and is externally evident as a saccular protrusion of variable size. Depending upon the structures involved, these defects are divided into three chief groups: (1) *myelomeningocele*, in which both the spinal cord and its membranes are included within the sac; (2) *meningocele*, in which only the spinal membranes compose the cyst; (3) *myelocystocele*, in which the spinal cord itself undergoes dilatation.

Myelomeningocele of the lumbo-sacral region is the most common form of spina bifida; much more rarely are the cervical and thoracic segments of the spine the seat of this defect. In rare cases the entire sacral region may be involved. Usually a rounded tumor, varying in size from a nut to an apple, appears in the midline about the juncture of the lumbar and sacral regions. When the child survives, it gradually increases and may attain the dimensions of a child's head.

The tumor is fluctuating, tense, and elastic, and often overhangs the lower margin of the cleft. When entirely covered by skin, the latter is attenuated and frequently cicatricial in appearance. More usually, however, the most prominent surface of the tumor is wanting in integument and invested by a smooth, soft, vascular membrane which is separated from the surrounding and elevated skin by a sharply defined boundary zone, covered with epidermis, and of a pale or bluish tint. The central part, uncovered by skin, is usually somewhat sunken and corresponds to the *area medullovasculosa;* the *zona epithelioserosa* and *zona dermatica* mark the intermediate and outer tracts of the tumor respectively. Pressure during labor, or the increasing tension following growth, may affect the nutrition of the integument and give rise to ulceration or gangrene, which results in perforation, destruction of the sac, or secondary cicatrices.

The defects of the skeleton include arrested development of the spinous processes and adjacent parts of the neural arches, the deficiency being most marked in the vertebræ contributing the lateral boundaries of the cleft. The latter may be limited to two, or even a single lumbar vertebræ; usually it involves the upper part of the sacrum, and may extend as far as the sacral fissure.

The spinal cord is commonly bent strongly backward and attached to the middle or upper part of the sac, the associated spinal nerves being correspondingly displaced. When the fissure lies near the lower end of the spine, the extremity of the cord may be drawn entirely into the sac, ending possibly in a flat expansion and without a conus. In many cases the cord extends lower in the spinal canal than normal. The pial investment is complete, and, in conjunction with the arachnoid, forms the sac enclosing the spinal cord; the dura, due to its imperfect development in association with the defective vertebræ, is always absent over the most prominent part of the tumor. In the much rarer and usually smaller thoracic and cervical examples of myelomeningocele, the spinal cord commonly protrudes less from the vertebral canal, being attached to the sac by a band-like fold. In exceptional instances the sac may escape through a ventral fissure, then constituting the rare defect *myelomeningocele anterior*.

Meningocele, in which the cyst is formed by the spinal membranes alone, occurs less frequently than sometimes assumed, since ordinarily the protruding membranes are closely associated with displaced nervous tissues. Uncomplicated spinal meningocele is encountered almost always at the lower end of the spine, the defects at the upper end of the column existing commonly in conjunction with hernia of the brain membranes.

The opening through which the meninges escape is not necessarily a posterior median cleft, since the sac may at times protrude between the arches, or through the intervertebral foramina or the sacral canal; rarely imperfections of the vertebral bodies allow the escape of the membranes anteriorly.

The sac of the meningocele appears oftener at the side, or downward, than in the midline. Since this defect is less unfavorable to prolonged life, the tumor not infrequently attains considerable dimension, at times increasing from the size of a hazelnut at birth to that of a man's head. The fluctuating cyst, filled with clear fluid, is covered with integument that may differ little from that of the surrounding parts; in other cases it may be tense and even gangrenous and rupture when subjected to unusual pressure. The wall of the sac consists principally of the abnormally distended arachnoidal supplemented by an imperfect dural layer which blends with the integument. The cavity of the cyst presents a smooth inner surface and communicates with the spinal subarachnoidal space; it is sometimes traversed by one or more spinal nerves, which otherwise run in the cyst wall, and may be secondarily subdivided into several compartments.

Myelocystocele or *syringomyelocele* implies a local dilatation of the central canal of the spinal cord, in consequence of which a larger or smaller segment of the cord, with the surrounding pia and arachnoid, becomes converted into a cystic tumor. The dura does not extend beyond the spinal canal. Since the sac represents the enormously dilated central canal, its lining consists of the ependyma cells and is, therefore, a layer of cylindrical epithelium. The remains of the compressed and atrophic nervous matter are seen on the ventral wall of the cyst as an area medullovasculosa to which rudimentary nerve roots may at times be traced.

Myelocystocele is often associated with lateral fissures and asymmetrical defects of the spine, especially shortening of the trunk. It may occur in any region, and is sometimes connected with anterior vertebral clefts.

When the dilatation of the cord is associated with distention of its membranes, the condition is termed *myelocystomeningocele*, a defect that may, when extensive, be mistaken for myelomeningocele. The differential diagnosis can be made by observing the character of the lining of the cyst, cylindrical cells being never found unless the sac is derived from the central canal of the cord.

Spina bifida occulta is the term applied to cases in which neither a cleft nor cystic tumor is externally visible. Such masked defects are usually of small extent and of sacral or lumbo-sacral origin. The position of these hidden imperfections is often indicated by a somewhat depressed or cicatricial area, or by a small cutaneous tumor. In other cases an unusual tuft of hair alone marks the location of the deeply seated defect, which on pressure may be detected as a minute opening in one or more vertebræ. In the cases carefully studied the greatly elongated spinal cord extended into the sacral canal and was connected with the external soft parts by a fibro-lipomatous mass. The spinal canal may, however, be dilated by the presence of such tumor, and the nerve roots forming the cauda equina may be unfavorably affected, resulting in considerable motor and sensory disturbance in one or both lower extremities. Paralytic club-foot and derangement of the functions of the bladder are among the consequences of these defects of the neural canal and its contents.

Doubling of the spinal cord (*diastematomyelia*) is an occasional accompaniment in rachischisis, more rarely in myelomeningocele; the duplicity consists in the cleavage of the immature cord, which is then represented by two usually atrophic or rudimentary bands, seldom by nervous cords of more perfect development. The presence of a thorn-like spur or process in certain cases suggests that the doubling of the cord may sometimes depend upon mechanical disturbance.

The *genesis* of spina bifida was formerly regarded as closely related to an accumulation of fluid within the spinal canal leading to rupture of the distended sac. Such origin of the defects under consideration is now generally considered untenable. The present views, however, are far from settled. According to v. Recklinghausen, whose classic investigations have greatly advanced our understanding of spinal clefts, the defective median union is occasioned by inadequate development, or aplasia, of the spine, due to deficient growth of the blastoderm as a primary cause. The observations of Tourneux and Martin, on the other hand, point to an incomplete separation between the medullary folds and the adjacent ectoderms as the immediate factor, in consequence of which the lips of the neural groove never become united. The surrounding mesoblastic tissue, the skleratome, in which the vertebræ develop, fails to produce arches which unite. Ziegler inclines to the view that the defective or arrested development is due to a "primary agenesis" affecting the germ, regarding the symmetry of the malformation as evidence opposed to the assumption of mechanical impressions from without. The marked differences in these defects as to the extent, situation, and details suggest the probability that their mode of production is by no means always the same. In complete rachischisis the rudimentary condition and incomplete separation of the medullary folds which preclude the closure of the neural groove not unlikely are associated with abnormality or excessive narrowness of the amniotic sac, whereby the arrested growth and nonunion of the medullary folds are favored. The explanation of the favorite location of myelomeningocele in the lumbo-sacral region is found in the fact that this part of the spine not only corresponds to the area in which closure of the neural canal is longest delayed, but is also subject to the disturbing influence of a too closely applied amnion, on account of the flexure which here is more pronounced than in the thoracic segment. The upper cervical region, occasionally the seat of spina bifida, is also involved in the marked flexure at the cephalo-cervical juncture. The observations of v. Recklinghausen have established the fact that in many cases, the length of the spinal cord is relatively excessive, a condition favorable to the production of kinks which at the points of least resistance may become additional factors in causing protrusion of the cord and its membranes. Hertwig, basing his conclusions on experimental studies on frog's ova, regards spina bifida as indicative of faulty closure of the blastopore, ascribing the arrest of development to influences which very early affect the germ-mass. That this malformation may arise apart from mechanical disturbance due to the amnion is shown by its occurrence in the *anamnia* (fishes and amphibians) in which fœtal membranes are never formed.

DEFECTS DUE TO ARRESTED DEVELOPMENT OF THE CRANIAL VAULT.—The defective formation of the vault of the skull presents a series of malformations which affect not only the brain case, but also the enclosed nervous mass. Since the arrest of development involves those parts of the skull which are analogous to the neural arches of the vertebræ, these malformations of the skull may be appropriately termed *cranioschisis*, of which two chief groups—*acranius* and *hemicranius*—are recognized; these depend upon the degree of the defect, and vary from total absence of the vault to partial arrest of development. The associated defective conditions of the brain mass include *anencephalus*, *exencephalus*, and *cephalocele*.

Acranius.—This malformation is distinguished by very slight or totally deficient development of the bones comprising the cranial vault, resulting in a skull in which the base is often the only part present. The brain shares in the defective development and is frequently wholly lacking, or at best represented by rudiments of nervous substance contained within a richly vascular spongy mass of connective tissue which forms a membranous covering for the base of the skull. This structure, the *area cerebrovasculosa*, corresponds to the area medullovasculosa already noted in connection with the defects of the spine, and consists of the remains of the pia and the nervous tissue.

Hemicrania implies a partial development of the vault bones, which, although producing more of the skull than present in acrania, results in an imperfect closure, principally in the posterior segment. This condition is often attended with protrusion of the brain substance (*exencephalus*) which may rise above the imperfect skull as a crowning mass or form a pendulous sac-like tumor over the occipital region. The protruding brain, although com-

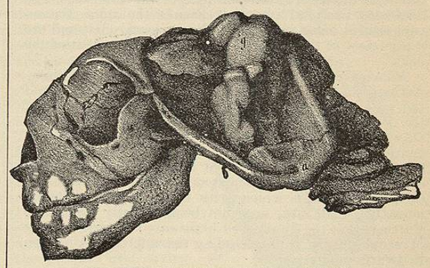

FIG. 4677.—Acranus Exencephalus. (Vrolik.)

monly uncovered by integument, is invested by a soft and yielding membrane representing the pia and arachnoid.

The defective development resulting in cranioschisis frequently affects also the adjacent cervical vertebræ, thereby giving rise to the condition known as *cranio-rachischisis*, in which a cleft extends from the posterior