



FIG. 4659.—Craniopagus Parasiticus. (Dömitz.)

find his brother dead. Within a little more than two hours of this discovery Eng's death quietly ensued, consciousness being retained almost to the last.



FIG. 4660.—Cephalo-thoracopagus with Janus-Head; other face presented malformed eyes, nose, and mouth. (Hirst and Piersol.)

The brothers were married when thirty-two years old and became fathers of large families, Eng having twelve and Chang ten children; of these twenty-two children all were normal except two of Chang's, who were deaf-mutes. Three years before Chang's death he was stricken with hemiplegia of the right side, from which he never entirely recovered. In January, 1874, Chang developed bronchitis which, aggravated by imprudent exposure, led to pulmonary oppression and inability to lie down without discomfort. After both had fallen asleep, Eng awakened to

The autopsy disclosed the bond of union to consist, in addition to the integument, connective tissue, fat, and aponeuroses, of a bridge of hepatic tissue, three inches long, one-half inch wide, and one-quarter inch thick, which arose on both sides, directly above the longitudinal fissure. Two peritoneal pouches on each side, extended transversely until separated by a vertical median septum. Extensions of muscular tissue of both diaphragms were represented by a broad slip from Chang, which was inserted into ensiform process

of Eng, and a similar but smaller band which passed to Chang, supplemented by two additional slips extending from Eng's diaphragm to that of Chang. The blood-vessels within the bond of union included a terminal and

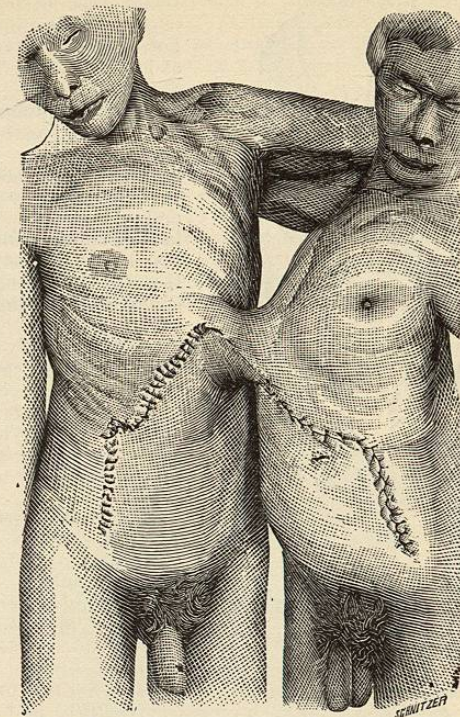


FIG. 4661.—Xiphopagus. "Siamese Twins." (Pancoast.)

an extrahepatic branch of the portal vein, branches of the hepatic artery, and some terminal twigs of the right internal mammary artery of Eng.

In this connection it may be of interest to note the operation undertaken by Prevost for the separation of the Brazilian xiphopagic sisters, "Rosalina" and "Maria." According to the letter published in the *Medi-*

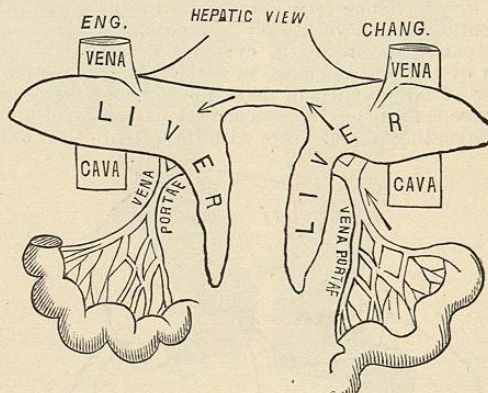


FIG. 4662.—The Hepatic Isthmus across the Connecting Band of "Siamese Twins" Eng and Chang.

cal Record (vol. lviii., 1900, p. 108) one sister, Rosalina, entirely recovered from the operation, the other succumbed.

Sternopagus implies an upward extension of the attachment, so that the sterna are fused into a single bone at the upper border of the common thorax. In cases in which the union is very intimate, the manubrium may receive the four clavicles of the two individuals.

The hearts may remain distinct, or they may be fused to form a single tubular organ, from either side of which the great vessels are given off. When two hearts are present they usually are enclosed within a single peri-

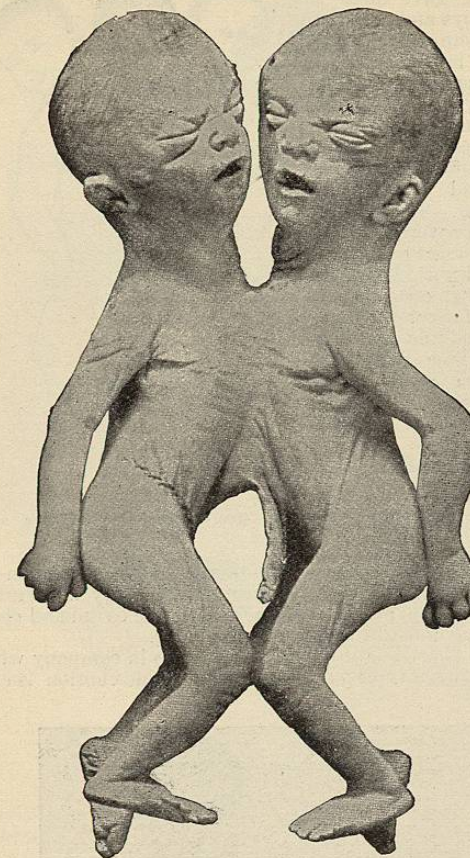


FIG. 4663.—Thoracopagus. (Hirst and Piersol.)

cardial sac. The common thoracic cavity is separated from the abdominal space by a single diaphragm. The middle portion of the digestive tube, including the greater part of the jejunum and ileum, is single. Above the jejunum, embracing the duodenum, the stomach and the esophagus, and likewise below the position of the vitelline duct, the digestive canal is double. The livers are blended, more or less, into one common mass. The genitalia are separate and distinct, each body having its own set.

While the limb complement of the sternopagic twins may be normal, deviations from a ventral toward a ventro-lateral type of union, due to convergence of the spinal axes, sometimes result in the blending of the approximated upper extremities, the third arm thus produced being the joint contribution of the two embryos. Such limbs are sometimes very rudimentary. Sternopagi are usually born dead; if alive, they survive only a short time in consequence of their defective heart development.

MODE OF PRODUCTION OF DOUBLE MONSTERS.—Apart from the more speculative considerations of the causes leading to double monsters which have already been briefly sketched, the manner in which duplicities are formed calls for notice. Attention has been directed to the two prominent opposed views—fission and fusion—which have prevailed, and the arguments have been reviewed that have influenced the adoption of the fusion theory as offering the most plausible explanation of the production of double monsters. With the exception of

the dicephali, in which dichotomous growth of the head end of the anlage may consistently be regarded as the mode of production, early union, more or less extensive, of the originally distinct but closely related, anlagen seems most worthy of acceptance as the principle upon which double monsters originate. The accompanying diagrams have been suggested by Marchand as hypothetically illustrating the most plausible genesis of the anterior and posterior types of duplicity. In Fig. 4664, A represents the assumed early stage in which the two closely related anlagen lie upon the common embryonic area, united to the chorion by the diverging belly stalks. With advancing development the latter become more widely separated, while the cephalic ends approach until they meet and union takes place; the umbilical vesicles meanwhile have become defined as two conspicuous sacs, one largely occupying the ventral surface of each embryo. The common amnion invests the dorso-lateral surfaces of both embryos and grows between their diverging belly stalks. C represents a later stage in which more marked fusion of the head ends has taken place, while at the same time the belly stalks are more widely separated, thus foreshadowing the later diomphalic or independent condition of the umbilical cords.

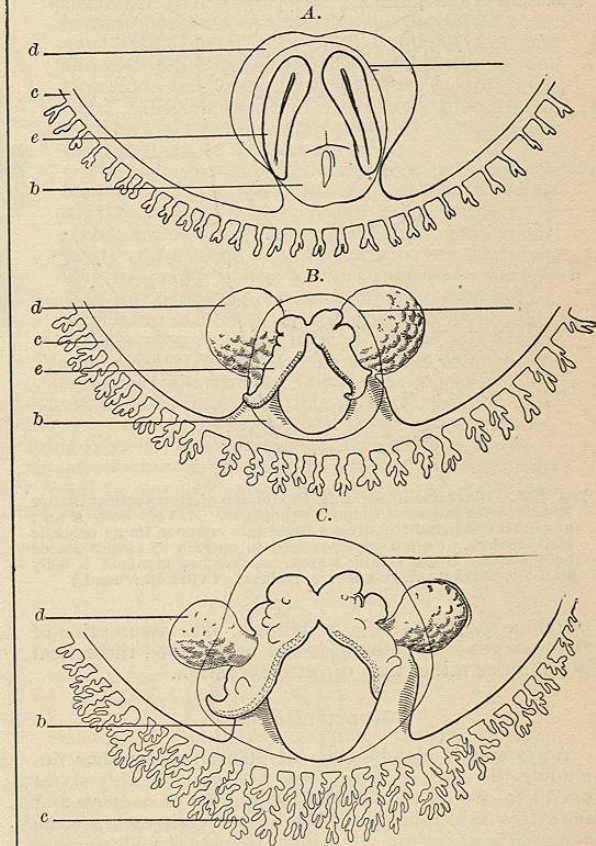


FIG. 4664.—Diagrams Illustrating Production of Hypothetical Double Monster with Anterior Union. Cephalopagus. A, The two embryos, on a common germinal area, are almost in contact with cephalic poles; united to chorion by closely placed belly stalks. B and C, later stages after union has been effected. a, Common amnion; b, belly-stalks; c, chorion; d, yolk-sac; e, embryo. (After Marchand.)

Rapid and extensive early separation of the belly stalks, together with rotation of the embryos, accounts for the various types of craniopagic twins.

In a similar manner the formation of the opposite condition of posterior union may be assumed to take place. (Fig. 4665.) The production of pygopagic twins, how-

ever, assumes the continued close relations of the belly stalk, with more or less divergence of the cephalic poles until, in certain cases, the axes of the two embryos correspond. The rotation and ventral type of union is associated with the retention of a common umbilical vesicle

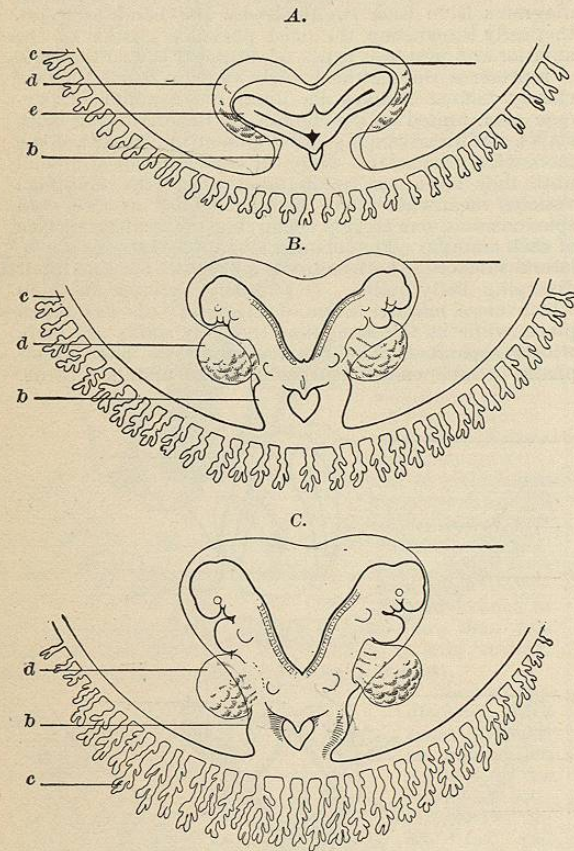


FIG. 4665.—Diagrams Illustrating Production of Hypothetical Double Monster with Posterior Union, Pygopagus. A, Very early stage; at caudal end primitive streaks fused into common tract; cephalic ends diverge. United embryos fixed to chorion by closely placed belly stalks. B and C later stages. a, Common amnion; b, belly stalk; c, chorion; d, yolk sac; e, embryo. (After Marchand.)

(vitelline sac) and single umbilicus. The assumption of these stages is hypothetical, and based upon theoretical possibilities rather than direct observation.

MULTIPLICITIES.

A systematic classification of pleural conceptions (including the cases of three, four, five, and possibly six at one birth) would call for the recognition of complete and incomplete multiplicities. The former again embrace the monochorionic group, of which the equal and unequal triplets are the best known representatives.

Monochorionic triplets are very infrequent, although a number have been described. The common chorion may enclose the three separate amniotic sacs, the umbilical vessels anastomosing within the placenta. Two authentic cases of monochorionic quadruplets have been recorded and an observation of five fetuses within a common chorion has been credited to Pigné. Other instances of quintuplets have been reported, but these, as in the case reported by Kölliker, include the association of monochorionic triplets with similar twins. The example of plural births reported by Vassali in 1888 recording the extraction of six fetuses, four males and two females, probably falls within this category. Kerr and Cook-

man have recently reported (*Medical Press and Circular*, May 27th, 1903) a remarkable case of six at one birth to which they were called. The mother, a native woman of Accra, West Africa, gave birth to five boys and one girl; all were alive when born, but succumbed between the second and fourth day. The girl and one boy had a placenta each, the remaining four children being attached to two placentæ. The mother, who recovered, exhibited a noteworthy predisposition toward pleural conceptions, three former pregnancies having yielded four, three, and three children respectively, making a total of sixteen in four confinements. The evidence sustaining a reputed instance of the birth of seven children, two boys and five girls, at Hameln in 1600, to which Barfurth⁶⁵ has called attention, is too uncertain to be convincing. Were the case satisfactorily authentic, here, too, the presence of more than one ovum and chorionic sac must be assumed as probable.

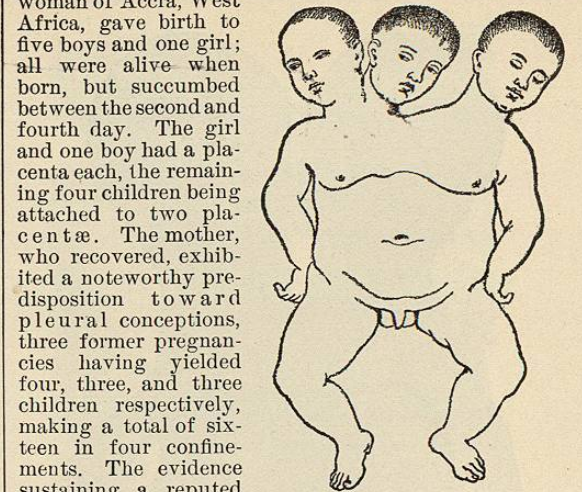


FIG. 4666.—Tricephalus. (Reina and Galvani.)

The occurrence of a malformed fetus in company with two or even three others within a single chorion is ex-

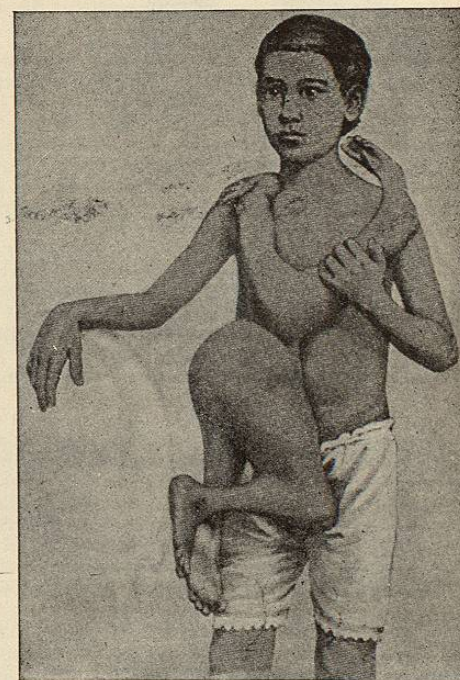


FIG. 4667.—Dipygus Parisiticus. "Laloo." (Hirst and Piersol.)

remely rare, only three trustworthy cases being recorded. That described by Soemmering presented four female fetuses within a common sac, three being normal while the fourth was an acephalus.

TRIPLE MONSTERS represent the rarest of all malformations. While a small number of such abnormalities have been observed among the lower animals, only a sin-

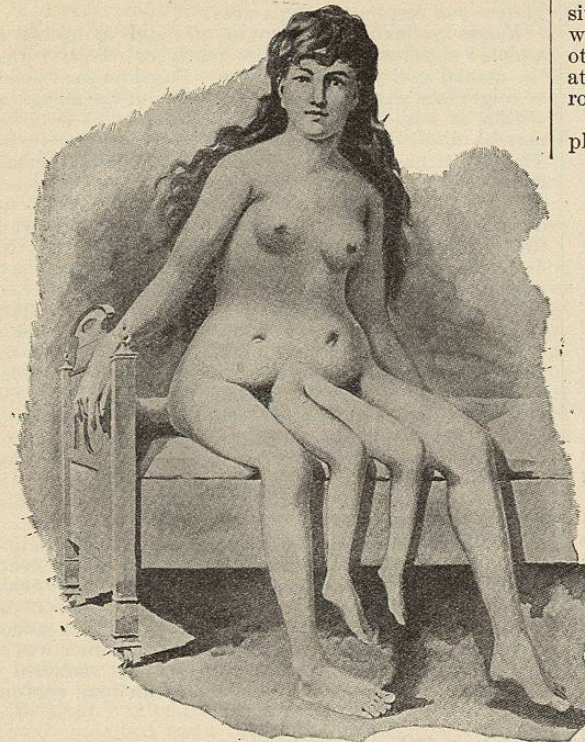


FIG. 4668.—Dipygus. (Hirst and Piersol.)

gle well-authenticated example of a human triple monster has been recorded by Reina and Galvani. The deformity was born in 1831, after a difficult labor lasting four days, necessitating the perforation and amputation of the first and second heads and the perforation of the third. Two spines were present bearing one and two heads respectively; the spinal columns were separate to their lower ends, although the sacra were very closely related. The pelvis included four iliac, two ischiac, and two pubic bones, the poorly developed medianly placed ilia being united by cartilage. The thoracic cavities were separated by a thin septum, each enclosing a heart. The abdominal cavity was large, the contained organs included a single stomach and an œsophagus that divided a short distance above its lower end; one of the resulting branches again divided, each head thus being provided with its own œsophagus. A similar arrangement was present in the trachea. The separation was apparent externally only on the neck and head, the unusually broad thorax seemingly being single. A third rudimentary upper extremity projected dorsally. The lower half of the body was normal, as were also the male genital organs.

The interpretation of the triplicity just described is to be found in the assumption that the anterior end of one of the primary anlagen, which have undergone posterior fusion in the production of the ischiopagus, has been affected by dichotomous growth. The result of the latter is seen in the closely associated heads surmounting one spine.

ASYMMETRICAL DUPLICITY.—Attention has been already called to the possibility that one of originally equal embryos which unite to form double monsters may suffer impairment of nutrition to such extent that its further imperfect development depends upon connections with the more fortunate mate. Such "parasites," there-

fore, result from secondary changes which affect embryos which were at first symmetrical. The continued nutrition and development of a part of the parasite, with the atrophy and disappearance of those parts less favorably situated, account for the remarkable cases in which a well-formed head is attached to that of the autosite. In other instances the cephalic end of one embryo becomes atrophic and is drawn within the body of the other, surrounded by the ventrally closing parietal plates.

Mention may here be made of two conspicuous examples of asymmetrical duplicity. The one, "Laloo"—familiar to many on account of his museum notoriety—represents the type of unequally developed thoracopagus, termed *dipygus parasiticus*, or gastro-thoracopagus *dipygus*. The "parasite," attached from the umbilicus to the xiphoid process of his host, exhibited neither voluntary movements nor responded to Laloo's will. An anus is not present, but a small penis occasionally voids small quantities of urine. The other malformation (Fig. 4668) is a remarkable instance of the rare form of posterior duplicity known as *diphygus*. In the case figured the duplicity begins at the third lumbar vertebra. She was married and became pregnant on the left side, both sets of organs being, however, functioning. Micturition and defecation take place independently on the two sides, but menstruation occurs at the same time. Anterior fusion of two closely placed converging embryonic anlagen probably accounts for this rare type of malformation.

The asymmetrical duplicities now to be considered include, on the contrary, the true parasitic double monsters which result from anlagen in which primary inequality and subsequent asymmetry of form and size are characteristic features. The inequality of the anlagen may be so marked that the parasite presents little or no external resemblance to a fetus; in other cases the dependent embryo may develop parts, as extremities, which are fairly well formed. In addition to such external attachments, or implantations, other parasites exist as inclusions within the autosite, having been drawn into the latter by the closure of the body walls, or lying from the earliest stages within the mesoderm.

As introductory to the consideration of the rudimentary fetal or parasitic growths encountered in various parts of the body, distinction must be made between der-

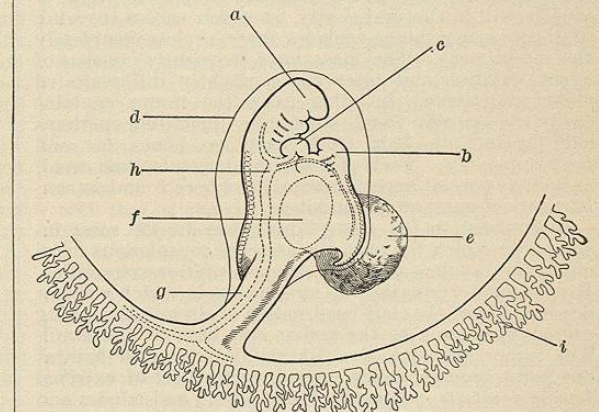


FIG. 4669.—Schematic Representation of Relations of Embryos of about Third Week in Production of Parasitic Duplicity. a, Auto-site; b, parasite; c, heart of former; d, common amnion joining common yolk sac (e) around f; g, common belly stalk; i, chorion. (After Marchand.)

moid cysts, arising from invagination and displacement of portions of the elements from the neighboring skin, and the true parasite tumors or *teratomata* originating by differentiation of tissues from the three germ layers de-

rived from the independent although imperfect development of a separate anlage. In the former case the tissues of the skin cyst include only those that ordinarily may

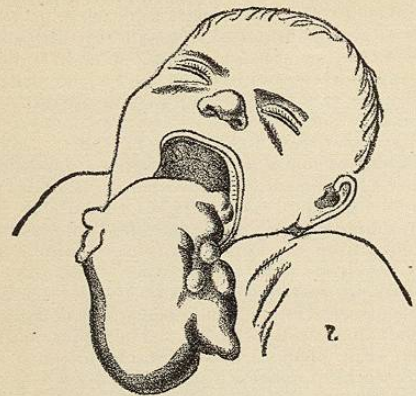


FIG. 4670.—Epignathus. (Ahlfeld.)

develop from the integumentary layers, their position being unusual owing to the dislocation of the aberrant cells ("Keimverirrung").

It is necessary, therefore, to recognize that simple dermoid cysts occur in many localities which may be the seats of true fetal growths. The distinction between the latter and the simple skin cysts is by no means always easily made. In doubtful cases, in which evident fetal structures are unrecognizable, the crucial test in establishing the parasitic nature of the growth is the presence of tissues and structures derived from all three germ layers, which cannot be attributed to transplantation of neighboring structures.

Although differing little in the principles governing their genesis, it is convenient for description to group parasitic duplicities according to the region of the autosite to which they are attached.

CEPHALIC PARASITES.—*Epignathus* is the term applied to a well-known group in which the rudimentary parasite is attached to the base of the skull or the palate and appears as a mass protruding from the mouth. The parasitic tumor is usually enclosed within an envelope of skin containing hair bulbs and sweat glands. When of small size, however, the tumor may lie almost entirely or wholly within the oral cavity, in which case a covering of mucous membrane replaces more or less completely the integument. The mass most frequently consists of cysts, cavities, and intervening slightly differentiated embryonal tissue. In other cases the tumor contains easily recognized fetal structures, including perhaps skin, epithelium, hair, teeth, cartilage, bones, fat, and brain substance. Parts of a rudimentary intestinal canal, as well as normal hepatic tissue, have been found as constituents of epignathic parasites.

In contrast to the amorphous types are the rarer instances in which the parasite presents conspicuous fetal characters, such as partly developed inferior extremities. Kreutzmann⁵⁶ described an epignathus in which a tumor depended from the cleft hard palate of the autosite, being united by a stalk to the vomer and base of the skull. The tumor, the size of a fist and covered with hair, at one point bore an imperfectly developed set of external female genitalia, possessing labia majora and minora and a tube representing the vagina. These lay between two rudimentary lower extremities.

As a great rarity the parasite may be attached to the region of the orbit from which a tumor-like mass protrudes. Ahlfeld⁵⁷ has described and figured (Fig. 4671) a remarkable case of this kind in which a buttock and left lower extremity protruded from the left orbit of a well-formed child. In the vicinity of the left oral angle projected an additional tumor. Broer and Weigert⁵⁸ also record a case in which the right eyeball was connected

with a tumor, the "size of an orange," that extended outward from the orbit. Microscopical examination revealed within the orbital fat the presence of cartilage, bone, epidermal masses, mucous gland cysts, portions of the digestive and respiratory apparatus.

Encranium embraces a group of fetal inclusions in which the parasitic anlage lies partly or wholly within the cranial cavity of the autosite. Based on their mode of origin (see page 693) and the position of the anlage, as within or without the skull, the intracranial parasites may be divided into *primary* and *secondary inclusions*. The former undergo but slight development and remain rudimentary, appearing as irregular tumors attached often in the vicinity of the sella turcica or beneath the dura in other localities. The immediate neighborhood of the pituitary body is a favorite location, not only for dermoid cysts derived from aberrant cells which have gained the interior of the skull by means of the early invagination producing the oral portion of the pituitary body, but also for growths which contain rudiments of various tissues and organs. Beck⁵⁹ found a tumor, the size of a walnut, on the sella turcica which contained cartilage, bone, myxomatous tissue, cyst lined with ciliated epithelium, and fourteen teeth; structure resembling thyroid gland was also present. Such tumors may remain for years without further growth, as in the case recorded by Eberth,⁶⁰ of a woman of seventy-five, in whom the teratome contained nervous, muscular, and lymphoid tissue.

It may be assumed that parasitic anlage, are drawn into the interior of the future skull during the early closure of the neural canal, to become later enclosed by the brain mass. Strassmann and Strecker⁶¹ described a teratoma in the right lateral ventricle which probably thus gained early access from the dorsal surface. The tumor was of the size of a walnut, riddled with spaces, contained a cyst lined with simple and stratified epithelium, nervous

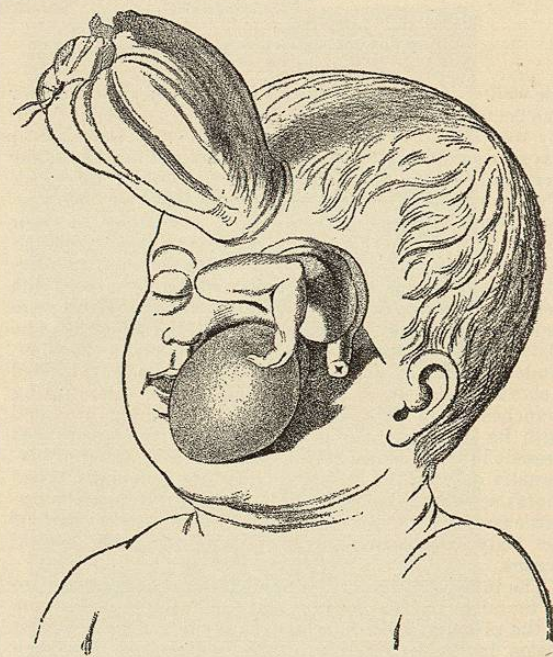


FIG. 4671.—Cephalic Parasite. (Ahlfeld.)

tissue, hyaline and fibrous cartilage, bone, adipose tissue, plain and striped muscle, lymphatic tissue, tubular and acinous glands.

Parasitic growths may be also attached to morphologically superior parts of the brain, as the pineal body. A pineal teratome, described by Gauderer,⁶² contained skin

and sebaceous glands, cartilage, bones, plain muscle, adipose tissue, and columnar epithelium.

The *secondary inclusions* may present externally fetal characters, such as more or less developed limbs attached to a rudimentary trunk. The escape of the inclusion may be through the base of the skull into or beyond the mouth, the protruding mass then resembling an epignathus. In rare instances the parasite may project from the cranium of the autosite. Aretæos⁶³ described a very remarkable case in which, in addition to a rudimentary inclusion, an acephalic parasite protruded from the skull of the autosite.

Lovett and Councilman⁶⁴ have also recorded an instance of multiple inclusions in a child of three weeks, which presented a scrotal and intracranial teratoma containing representations of all three blastodermic layers.

The encranial parasites above described imply the production of more or less characteristic fetal structures from the development of independent anlages capable of giving rise to all types of tissue. The inclusions from such anlages may, therefore, be regarded as true parasites, although they may remain rudimentary. To be distinguished from the latter are the intracranial dermoid cysts originating from the isolated and transplanted masses of epidermal elements, which ordinarily produce the prototypes of the tissues found within the intracranial growths. The latter should not be included within the true parasitic inclusions.

CERVICAL PARASITES OR TERATOMATA comprise the true fetal inclusions attached to some part of the region formed by the visceral arches. Such parasitic growths appear as cystic tumors, covered with integument and extending a variable distance along the neck and lower jaw. In addition to the cysts usually present, the tumors contain more or less developed fetal structures, including bones, teeth, parts of the head, and extremities. A cervical teratome in a child of two years, described by Brunker,⁶⁵ extended from the zygoma to the third rib and contained brain tissue, fat, cartilage, striated muscle, glands, and a rudimentary eye. Usually the seat of the growth is less extensive and is limited to the lateral cervical region. Instances are recorded of attachment to the parotid and the oral region, the latter cases forming a transition between the cervical and epignathic groups. Gurlt and Slomann both have described cases in which teeth were conspicuous; in one instance in a child of five years, both milk and permanent teeth were present and corresponded in form and size to the age of the patient.

THORACIC PARASITES.—Closely associated with the teratomata of the cervical region are the dermoid growths occasionally encountered within the anterior mediastinum in the vicinity of the thymus gland or its remains. That the migration of this organ from its original relation with the third pharyngeal pouch to the mediastinal space is the probable means of transplantation of the primary tissues giving rise to the dermoids under consideration is suggested by the intimate relations between the outer and inner germ layers in the occluding membranes closing the visceral clefts in mammals. Although the primary thymus anlage is entodermic in origin, the proximity of the ectoderm lining the external furrows and the important changes in the relation of the arches effected by the formation, and later closure of the sinus precervicalis render the transportation of ectoblastic and mesodermic tissue in connection with the migration of the thymus highly probable. Additional evidence of close association is had in the presence of thymus tissue in cases reported by Marchand and by Pinders.

As shown by the series of forty cases tabulated by Christian,⁶⁶ the great majority of mediastinal dermoids are to be regarded as tumors of ectodermal origin, since of these cases only two can be classified as true teratomata, the derivatives of all three germ layers. These conclusions agree with the results of the studies of Pflanz⁶⁷ and of Wilms.⁶⁸ A few cases, however, such as recorded by Virchow,⁶⁹ Marchand,⁷⁰ Ekehorn,⁷¹ and Fletcher,⁷² give evidence of being true teratomata. The

possibility of other congenital tumors of the lower anterior mediastinum being the result of inclusions at the time of the closure of the ventral body walls cannot be ignored, although a branchiogenetic origin is undoubtedly the more common. Extensions to the neighboring lung constitute the pulmonary dermoids sometimes described.

Although among the lower animals examples of parasitic anlages attached to the dorsal body surface are occasionally observed as rudimentary limbs projecting from

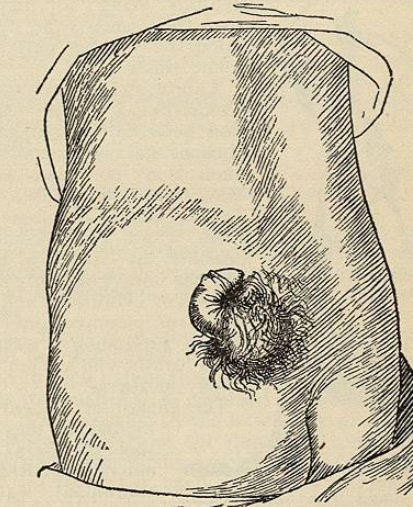


FIG. 4672.—Supernumerary Penis on Back. (Windle.)

the back, such malformations have not been encountered in the human subject. In this connection mention should be made of the remarkable case (Fig. 4672) described by Windle⁷³ of a young man who, in addition to normal genitalia, possessed over the lumbar vertebrae an elevation similar to a mons from which projected a small but well-formed penis, capable of erection, although imperforate. Windle refers to a somewhat similar case, recorded by Tsortis, of a soldier of twenty-one, who bore, below the left scapula, an almost complete set of female external genital organs. A small cavity between the labia resembled a rudimentary vagina, above which was a clitoris-like tubercle. These cases can be explained only on the assumption of the partial development of a second, originally independent embryonic anlage.

SACRAL PARASITES AND TERATOMATA.—Of all forms of parasitic inclusions those occurring in the sacral and perineal regions are most frequent. They usually appear as cystic tumors attached to the sacrum, on either the anterior or posterior surface, or to the coccyx. The tumor is covered with integument directly continuous with that of the autosite, and consists usually of several cysts. Not infrequently the position of the teratome is not median, but to one side; in such cases often a second enlargement, consisting of a simple cyst, lies at the side of the larger nodulated parasitic growth, the two tumors sometimes occupying the opposite sides of the coccyx. Within the compartments of the teratome are found more or less striking evidences of its fetal origin. These may include various epidermal structures and connective tissue, cartilage, bone, muscle, and nervous tissue, as well as parts of the spine, the digestive tract, and glands. In exceptional cases rudimentary extremities, both upper and lower, may be present and even free, being dependent from the tumor. In the classic and unique case of Anna Przenosly described by Ammon, in the third year a small congenital tumor opened and revealed a supernumerary leg which after seventeen years was removed by amputation. The presence of considerable masses of muscular tissue within the sac surrounding the inclusion may give rise to movements as in the