formations. Its development may be imperfect, resulting in retention of the fœtal tubular form and under-size, the stomach at times being little larger than the succeeding gut. The organ may present constrictions, subdivisions, and dilatations. Sometimes the pyloric orifice may be partially or entirely closed, the stenosis being occasionally seen in infants. In company with other abdominal viscera, the stomach may suffer displacement in large congenital umbilical herniæ, as well as transposition in situs inversus. Total absence of the stomach may occur in acephalus.

The Intestines.—Partial persistence of the vitelline or omphalo-enteric duct in the form of *Meckel's diverticulum* is the most frequent congenital deformity of the intestinal tract. The duct for a time normally is pervious, but before birth it loses its lumen and is represented by an atrophic fibrous cord passing from the umbilicus to the ileum. When retained, the resulting diverticulum opens into the ileum, usually a little over a metre from the ileocolic valve. Its length varies from a mere shallow recess to the entire distance from the gut to the umbilicus; usually, however, it is some 10–13 cm. Its width at the intestine is commonly that of the gut, the blind extremity being somewhat less. The latter may exhibit a pouched condition, or a rounded end similar to the finger of a glove. The diverticulum usually springs from the convex side of the intestine, but may join the latter obliquely, or on the side of the attachment of the mesentery. In the latter case the diverticulum is provided with a process of mesentery. In exceptional cases the duct retains its lumen throughout, and on the disappearance of the stump of the cord after birth opens at the umbilicus, thus establishing a congenital umbilical *fistula*. Sometimes a small portion of the pervious duct remains attached to the umbilicus and becomes the basis of a tumor-like mass, which may yield a scanty secretion derived from the modified mucous membrane of the vitelline duct. The diverticulum may become constricted and its communication with the gut entirely lost, it remaining attached to the intestine as a cyst-like appendage. The latter may mark the seat of active growth resulting in the production of an intestinal cyst of huge size.

Total absence or extensive defects of the intestinal tube are always associated with grave general malformation, as in acardiac monsters.

Contraction and *closure* of the lumen of the intestinal tube may occur at different points, or a portion of the

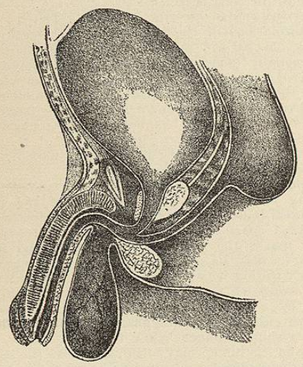

Fig. 4683.—Atresia Ani with Rectum opening into Urethra. (Witt.)

gut may suffer obliteration. The upper part of the duodenum, in the vicinity of the entrance of the bile and pancreatic ducts, is a favorite position for interruption, the atresia of the gut being at times followed by extensive dilatation of the beginning of the duodenum in which the stomach may also be involved. The observation of Tandler,[97] that temporary constriction, or even occlusion, of the gut occurs near the entrance of the com-

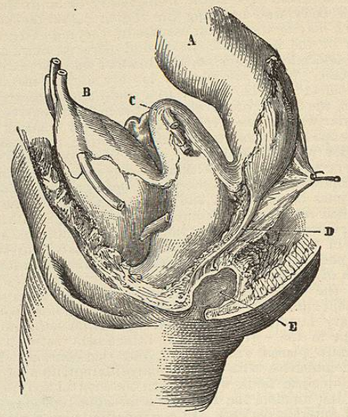

Fig. 4684.—Atresia of Rectum. *A*, Rectum connected with anal canal (*E*) by means of fibromuscular cord (*D*). (Guinard.)

mon bile duct during normal development of the duodenum, is suggestive in connection with atresia of this portion of the intestine. Ratz [98] reports an interesting case in which the small gut ended blindly, followed by the closed beginning of the large intestine. No evidences of inflammatory processes were present. The cæcum and appendix were wanting.

Malformations of the *lowest segment* of the gut include an interesting group of defects sometimes dependent upon the faulty development of the parts concerned in forming the outlets of the intestinal and genito-urinary tracts. Reference to the early relation of these parts recalls that for a time a common space, the cloaca, receives the gut dorsally and the urogenital sinus ventrally. Separation of these tracts is effected by the extension of the urethro-rectal septum, which grows downward until it meets and fuses with the cloacal membrane closing the cloaca. This structure, the remains of the primary anal membrane, consists of the opposed entoblast and ectoblast and forms a temporary wall between the cloaca and the exterior of the body. Independent openings for the urogenital and intestinal canals are secured by the breaking down of the epithelial layers constituting the occluding membrane.

A conspicuous defect directly referable to arrested development is *persistence of the cloaca*, in which the rectum and urogenital sinus retain their primary relation, in consequence of the failure of the septum to appear, and open into a common space. The latter, closed by the still existing cloacal membrane, becomes distended by the accumulation of the excreta from the intestinal and urinary tracts. More usually the separation of the urogenital sinus and the gut is partially accomplished, the imperfection in the septum being represented by a permanent communication between the two canals; hence in the male the rectum opens into the urethra (usually the membranous but sometimes penile portion), or, rarely, into the bladder (*atresia ani vesicalis*). Occasionally the rectum empties into the vagina, very exceptionally into the uterus.

Imperforate anus (*atresia ani*) is relatively frequent and depends upon more or less extensive arrest of development of the gut, coupled with secondary attach-

ments, which lead to the formation of a fibrous septum, closing the anus. The rectum may be well formed, or so defective (*atresia recti*) as to end blindly at some distance above the position of the anus, lying free and distended within the pelvic cavity. The atrophied gut may be attached by a fibrous cord without lumen. On the other hand, a well-developed anus may exist with closure of the rectum. In exceptional cases of imperforate ani, the rectal opening or fistula may lie farther forward and be found on the perineum, scrotum, or along the penis. The fistula may, on the other hand, be found over the sacral region, the unusual situation being probably associated with the primary dorsal position of the earliest trace of the anal depression, which under usual conditions acquires a more ventral location.

Variations in the form and situation of the *large intestine*, especially the colon and the cæcum, are very common, since the position and relations of this part of the intestine are greatly influenced by the torsion and secondary growth of the early gut tube. With the exception of stenosis, atresia, and possibly dilatation, the congenital defects are variations rather than malformations. Associated with these variations, modifications are also encountered in the arrangement of the peritoneum. The most striking of these is the retention of the fœtal simplicity of the mesentery (*mesenterum commune*), in which the duplicature enclosing the small intestine is directly continuous with the mesocolon and uncomplicated, the usual overhanging and anterior position of the large gut being absent, owing to the upward displacement of the latter having never occurred.

The *liver* is only seldom the seat of malformations, unless associated with general defects involving the intestinal canal, as in acephalic monsters, in which the organ is wanting. Very rarely absence of the liver may be unaccompanied by other malformations. The more usual, but still infrequent, defects include abnormalities in the size and number of the lobes; accessory or dislocated lobes may occur within the falciform ligament. Absence of the gall-bladder has been observed by Marchand in a child otherwise normal. Congenital stenosis of the bile passages, or enormous distention of the common duct due to obstruction near its termination in the duodenum, as well as unusual situations for the latter, are also occasionally encountered. Displacement and modified form are commonly seen in connection with umbilical and diaphragmatic herniæ or abdominal clefts.

The *pancreas* shares with the intestinal tube the effect of profound general malformation involving the alimentary canal. Supernumerary or accessory glands occasionally lie embedded within the intestinal, or at times gastric, wall. They may remain of small size and hidden, or they may become later in life the seat of cysts of large size. Unusual arrangement and abnormally situated openings of the pancreatic duct are dependent upon variations and connections in the development of the original anlages. The pancreas may suffer displacement in consequence of abnormal relations of the duodenum and stomach, or it may share the general inversion accompanying transposition of the viscera.

As a matter of convenience, the congenital defects of the spleen and of the suprarenal body may be here introduced, these organs having, of course, no connection with the digestive apparatus.

The Spleen.—Very rarely the spleen is entirely wanting. Variations in the size of the normal organ include such a wide range that it is difficult to establish the limit of pathological defect. The most common malformation is the presence of one or more accessory spleens, which are usually found in the immediate vicinity of the chief organ, although they may lie at some distance or within the pancreas. H. Albrecht [99] has described a remarkable case of multiple spleens, in which a principal organ in the usual position was wanting and instead segments or supernumerary bodies to the number of almost four hundred were scattered throughout the abdomen. They occurred in groups which occupied the parietal peritoneum, the right half of the diaphragm, the upper surface and the falciform and coronary ligaments of the liver, the root of the mesentery, the splenic flexure of the colon, and the anterior surface of the rectum within the pouch of Douglas. Histological examination confirmed the macroscopic diagnosis of multiple accessory spleens. It is highly probable, however, that in many cases the supposed numerous minute accessory spleens are really hæmolymph glands of the splenic type (Warthin).

The Suprarenal Body.—The adrenals may rarely be wanting, or abnormally small, such defect being usually associated with deficient brain development (anencephalus). This relation, although recognized by the older writers, has been emphasized by Zander,[100] according to whom the development of the suprarenal suffers when the anterior half of the cerebrum is rudimentary; neither deficiency of the posterior part of the cerebral hemispheres and brain stem nor cerebral atrophy after the suprarenal has developed produces diminution of the latter.

The most frequent abnormality is the occurrence of accessory suprarenal bodies. The structures formerly described under this name included not only those connected with the adrenal or located in its more immediate neighborhood, as in the kidney or the liver, but also those found at a distance in the vicinity of the sexual glands, as in the broad ligaments or between the testicle and epididymis. The careful studies of Aichel [101] have shown that the last group represents constant organs of normal occurrence (*superadrenals*), which develop in relation with the atrophic tubules of the Wolffian body, and that the designation "accessory" is properly restricted to the supernumerary bodies which probably arise from constriction and isolation of a portion of the suprarenal anlage.

Malformations of the Respiratory Tract.—The *nose*, in addition to the defects associated with facial clefts, may present congenital malformations involving defective development of the septum, ethmoid, nasal, and turbinated bones. Narrowing and closure of the posterior nares also occur.

The *larynx*, in common with other parts of the respiratory tract, may be entirely wanting, such conditions, however, being associated with grave malformations (acephalus) profoundly affecting the alimentary canal from which the pulmonary apparatus is the direct outgrowth. The congenital defects of the larynx consist usually of abnormalities affecting the cartilages, which may be increased in number, as a doubled epiglottis, or of the ventricle, which may be of unusual size with exaggeration of the lateral laryngeal pouch, the latter at times extending as far as the level of the hyoid bone and suggesting the laryngeal sacs seen in anthropoid apes. Abnormalities of the larynx as to size, excessive or deficient, as well as marked asymmetry, are occasionally observed. Complete closure of the larynx has also been recorded.

The *trachea* at times is absent, the respiratory tube undergoing bifurcation into the bronchi immediately below the larynx, or it may be only abnormally short. Contraction and occlusion also occur. Communication with the œsophagus has already been noted in relation to the defects of the gullet. The remains of such fistula may become a cyst connected with the posterior wall of the wind pipe. The cartilaginous rings are subject to variations in number and size in consequence of division or of fusion. Abnormal branching of the trachea into three bronchi is occasionally seen, the additional bronchus usually passes to the right upper lobe and is regarded by some (Chiari) as a supernumerary eparterial twig springing from the trachea instead of from the right bronchus. Herxheimer inclines to the view that the variation indicates the formation of an accessory lung. This author [102] has recorded an instance in which the first division of the trachea occurred 2.5 cm. below the cricoid cartilage, the right tube passing to a small nodule that presented all the histological characteristics of functionating pulmonary tissue. Rarely an additional eparterial or apical bronchus occurs on the left side in connection with a

left lung possessing a third lobe (Chiari). Rudiments of supernumerary bronchi sometimes give rise to congenital diverticula.

The *lungs* are relatively infrequently the seat of primary congenital malformations, although the entire organ on one side may permanently remain uninflated and rudimentary. Such condition may be induced by the intrusion of abdominal viscera which have migrated into the thorax through a diaphragmatic hernia. More usually the defective development of the pulmonary tissue is limited to one part of the organ, the affected area consisting of branches of the bronchial tree surrounded by a relatively dense mass composed chiefly of highly vascular tissue without alveoli, the conversion of the primary epithelial bud-like divisions of the forming organ having never taken place. The bronchial tubes within the rudimentary area may appear normal, but they sometimes exhibit great dilatation and may be converted into congenital bronchial cysts, which may occupy a large part of the entire undeveloped area.

Variations in the number and size of the lobes are frequent, especially in the left lung, in which three lobes are not rarely seen. The division, according to Fürst, is commonly due to the presence of an unusual pleural fold, which effects cleavage of the developing lung. In certain instances the additional lobes may possess morphological interest, as pointed out by Schaffner, in connection with the right inferior accessory lobe, which he regards as the homologue of the constant cardiac lobe seen in other animals.

Rarely a rudimentary supernumerary or accessory lung, lying between the normal organ and the diaphragm, is formed in consequence either of an early division of the original anlage and isolation of the separated portion, or of the outgrowth of an additional independent anlage from the primitive gut tube. An interesting example probably involving such latter condition was recorded by Wechsberg.[103] The rudimentary lung was represented by an ovoid tumor, 5 cm. in its longest diameter, that projected into the left pleural sac, and was attached to the œsophagus by a thin stalk, about 2 mm. above the diaphragm. An aberrant course of the arching vena azygos major and the production of an unusual fold of parietal pleura have been held accountable for the presence of an isolated apical lobe in the right lung (Gruber).

Malformations of the Circulatory Organs.—Under this heading will be considered only the congenital defects of the heart and of the closely associated large blood-vessels, the discussion of the variations affecting the smaller vessels belonging less to the domain of teratology than to that of anatomy.

Malformations of the Heart.—Since congenital defects of the heart largely result from arrested or perverted progress of development, a brief review of the salient features in the formation of this organ will aid in appreciating the significance of the malformations described.

After the fusion of the two, at first widely separated, anlages to form the single straight heart tube, the flexion and torsion of the latter results in the production of a flattened S-like tube, of which the antero-inferior portion is the arterial and the postero-superior are the venous segments. These two divisions soon become partially separated by an external groove, which indicates the position of the auricular canal, the primitive auriculo-ventricular opening by which they communicate. The superior continuation of the arterial segment constitutes the truncus arteriosus. The subsequent conversion of the primary two-chambered heart into one, possessing right and left compartments, and of the truncus arteriosus into the aorta and the pulmonary artery is accomplished by the formation of a longitudinal partition consisting of three parts. 1. An interauricular septum, which, beginning in the fourth week on the upper and hind wall of the primitive auricle, grows downward toward the orifice into the ventricle and separates the original venous chamber into a right and a left half. This septum, however, is for a time incomplete, since until after birth it contains the foramen ovale, which permits the blood stream to pass directly from the right into the left auricle. 2. An interventricular septum, which, starting below, gradually effects the division of the common ventricular cavity into a right and a left ventricle. The primary auriculo-ventricular orifice likewise becomes differentiated into a right and a left division by the formation of the septum intermedium, produced by the downward growth of the interauricular partition, and its fusion with the thickened margins of the auricular canal. The separation of the two ventricles is completed last in the upper part of the interventricular septum, where for a time communication between the two sides persists. 3. An arterial septum, which starting some distance above, extends downward toward the heart to meet the interventricular partition and divide the truncus arteriosus into the pulmonary artery and the aorta, which then communicate with the right and left ventricles respectively. The primary disposition of these vessels is such that the aorta lies directly in front of the pulmonary artery; their later relations to each other and to the heart chambers are acquired secondarily in consequence of the rotation and torsion to which they are subjected during development.

Malformations of the septum constitute the most frequent congenital defects of the heart; of these the imperfections of the *interauricular* partition are most common. That such should be the case is to be expected, since the existence of the foramen ovale predisposes to defects of the septum. The *persistence of the foramen ovale* may be complete, with correspondingly large opening; more usually partial closure has taken place, leaving sometimes only a mere cleft between the anterior margin of the foramen and its valve. Fawcett and Blackford[104] conclude that faulty closure of the foramen ovale exists in 28.3 per cent., the opening (most common in female subjects) varying from 1 to 15 mm. A much rarer defect at times occurs at the lower part of the interauricular partition due to faulty union between the latter and the lips of the primitive aural canal. *Total deficiency* of the interauricular septum, a three-chambered heart resulting, occurs very rarely and then in conjunction with other grave defects.

The *interventricular septum* is the seat of defects much less frequently than is the wall separating the auricles. When present, these malformations usually occupy the upper and anterior part of the septum, known as the pars membranacea. This corresponds to the position in which closure is last effected by union of the upward growing crescentic inferior fold, the septum intermedium and the partition dividing the truncus arteriosus into aorta and pulmonary artery. Since the last-named partition is concerned in completing the interventricular wall, imperfections in the latter are not infrequently associated with malposition of the former. This condition may result in disproportionate division of the lower end of the truncus arteriosus, the aortic orifice being enlarged at the expense of that of the pulmonary artery. Complete absence of the interventricular septum may occur, or even failure of both the auricular and ventricular parts of the general longitudinal partition, the heart under such conditions containing but two chambers.

Malformations of the great vessels are connected with the imperfect or perverted development of the septum, which is formed within the truncus arteriosus by the union of two elevations from opposite sides of the vessel. Normally the equal subdivision of the latter vessel is insured by the median position of the partition. Deviations from this by the ridges forming nearer one wall than the other evidently must lead to disproportion in the size of the resulting vessels, the aorta and the pulmonary artery. Such defects are frequently associated with imperfections of the interventricular partition, since the septum within the truncus takes part in the division of the ventricular chamber by joining the upward growing fold. Sometimes, however, extensive deficiency of the aortico-pulmonary septum may exist with normal valves, as in the case described by Hektoen.[105]

Abnormalities in the *number of the leaflets* of the aortic and pulmonary semilunar valves include both increase and decrease. Such congenital variations are usually referable to atypical division of the four primary leaflets which guard the lower end of the truncus arteriosus. When separation of the latter into two new vessels occurs, each elongated lateral primary leaflet is subdivided, of the six resulting segments three going to form the aortic and the pulmonary valve each. Fusion and cleavage of the original segments are responsible for diminution and increase of the leaflets guarding these openings, which may be so few as two or so many as five. The variations produced by pathological processes affecting the valves are manifestly beyond the present consideration.

Stenosis of the pulmonary artery, commonly coupled with narrowing or closure of its cardiac orifice with defective semilunar valves and possibly contracted conus arteriosus, is not infrequent. Although this malformation may exist unaccompanied with a defective interventricular septum, it frequently occurs associated with the latter condition. Deflection of the septum of the truncus may result in abnormal origin of the great vessels, both being connected with the right ventricle, or transposition of their usual relations occurring, the aorta springing from the right ventricle and the pulmonary artery from the left.

Stenosis of the ascending aorta with excessive size of the pulmonary artery sometimes exists, but is far less common than the opposite condition. Such narrowing may likewise occur independently or in conjunction with septal and other defects. Local constriction, or even complete closure and obliteration, at times takes place, as at the "isthmus" between the left subclavian artery and the ductus arteriosus, in consequence of which the circulation is maintained by the enlarged collateral vessels.

Persistence of the ductus arteriosus may occur without other defect, or it may be a compensatory retention, as when the narrowing of the pulmonary orifice exists to a degree that renders necessary the persistence of the vessel as the means of maintaining an adequate pulmonic circulation, the lungs being supplied by the blood that passes into the pulmonary arteries from the aorta by way of the ductus arteriosus. Stenosis of the aortic valve when of high degree is usually associated with feeble development of the left side of the heart and persistence of both the ductus arteriosus and the foramen ovale. By these channels the blood forced into the pulmonary artery partly passes by the ductus into the aorta and general circulation, while the foramen ovale provides the means by which the blood returned from the lungs may escape into the right auricle to mingle with that eventually propelled from the right ventricle.

The *auriculo-ventricular valves* may also present variations involving the number of the segments and the size of the opening. Such defects depend upon abnormal position, fusion or cleavage of the pad-like thickenings, and elevations of the margins of the orifices from which the leaflets are formed. Congenital narrowing, closure or obliteration of the auriculo-ventricular apertures may occur in consequence of fusion of the early segments, or of deflection and union of the septum intermedium with one side of the opening. When closure of the latter is complete, the maintenance of the circulation is possible only when a defective septum exists.

Abnormalities in the number and arrangement of the papillary muscles, the presence of unusual bands, the excessive prominence of the columnæ carneæ, the honeycombed myocardium, etc., depend upon faulty development and consolidation of the muscular heart, which at first consists of a network of contractile trabeculæ.

Variations in the openings of the venous trunks emptying into the right auricle also occur and are dependent upon arrested or disturbed development of the sinus venosus. The latter vessel, the common chamber for the reception of the venous trunks returning the blood to the heart, communicates with the primitive auricle by a single large opening guarded by two valves, a right and left. With the growth and expansion of the heart, the sinus venosus loses its independence and is drawn into the auricle, of which it constitutes the posterior portion. In consequence of this change, the tributaries of the sinus now open by separate apertures directly into the right auricle as the two cavæ and the coronary sinus. Should arrested development prevent the absorption of the sinus, the venous trunks may communicate with the auricle by a common orifice. Stenosis and atresia of these apertures sometimes are observed. The thread-like trabeculæ, at times forming networks, which are occasionally attached to the Eustachian and Thebesian valves, represent, according to Chiari, the remains of the septum spurium and the right valve, which originally guarded the entrance of the sinus venosus.

Variations in the manner in which the *pulmonary veins* terminate in the left auricle are explained by the early relation of these vessels to the heart. The four pulmonary veins at first join to form a single trunk, which empties into the left side of the original auricular division of the young heart before the organ is separated into a right and left half. With the growth and expansion of the auricle, the short single vessel disappears in the heart wall and the separate pulmonary veins open into the auricle by independent openings. Where the extra-cardiac union is more extensive than usual, or the absorption of the primary single trunk is imperfect, the early relations persist, resulting in a simple, double, or triple venous opening.

The *large arterial trunks* arising from the arch of the aorta frequently present variations in their origin and course, which are referable to abnormal metamorphosis of the five pairs of aortic arches given off from the ventral continuation of the truncus arteriosus. The detailed consideration of these anomalies, which especially involve the innominate, common carotid, and subclavian arteries, belong to descriptive anatomy rather than to teratology. The rare occurrence of double aortæ is to be referred to the persistence of the two trunks of early embryonic life.

The *great venous trunks* entering the heart are subject to abnormalities which depend upon either defective development of vessels formed secondarily, or persistence of primary vessels that usually disappear, or at most are of small size. Results of the first cause are seen in the absence of the inferior vena cava above the renal veins, of the left innominate and of the left common iliac vein.

Examples of abnormal persistence are encountered in the presence of the left cardinal veins and of a left duct of Cuvier in the form of exaggerated hemiazygos veins and a left superior cava respectively. Very usually the retention and excessive size of primary veins which ordinarily atrophy are compensatory and due to the defective development or absence of important secondary trunks.

Malposition or transposition of the heart (*dextro-cardia*), in which the relations of the organ and its great blood-vessels are accurately preserved in reversed order, is usually associated with general situs inversus involving all the viscera within the body cavities. This condition has been already considered. Very rarely the transposition affects the heart and great vessels alone, the remaining thoracic and abdominal organs retaining their normal positions. Reference has been previously made to the partial transposition which may involve the aorta and pulmonary artery and the ventricles in consequence of faulty division and torsion of the truncus arteriosus. Durante[106] describes a striking example of this malposition. The displacement which the heart suffers in conjunction with extensive clefts of the ventral body wall may result in an extra-thoracic position of that organ (*ectopia cordis*). Doubling of the heart has been described as a very rare malformation. Its possible occurrence is ascribed to imperfect fusion and continued independence of the two primary heart tubes, which ordinarily blend to form a single organ.

Malformations of the Uro-Genital Organs.—*The Kidney.*—In rare cases and often coupled with other grave defects, both kidneys may be totally wanting, or

so rudimentary as to be practically absent, being represented by only meagre suggestions of renal tissue. Dykerhoff[107] has called attention to the fact, and described an illustrative case, that living and otherwise well-developed children are sometimes born with aplasia of high degree of both kidneys and obliteration of the ureters. Such defects evidently lead to death within a short time after birth. Since the epithelial elements of the organ are derived as expansions of the primitive ureter, which in turn is an outgrowth from the Wolffian duct, the rudimentary condition of the kidney is usually associated with imperfect development of its excretory duct. More frequently only one kidney, most often the left, is rudimentary, in which case congenital compensatory hypertrophy of the one present exists. Deficiency of both organs is manifestly incompatible with life.

The commoner malformations of the kidneys include various degrees of *fusion* and *displacement*. Union of the two organs at their lower ends produces the *horseshoe kidney* often observed. The connection varies in position, size, and intimacy, in some cases the union being little more than apposition; in others a fibrous band forms the junction, while at times actual fusion of the renal tissue takes place. Much less commonly the intervening bridge may occupy the upper or middle portion of the adjacent surface, or the entire opposed borders of the two organs may be united. Two separate pelves and ureters are usually present, the latter sometimes crossing the anterior surface.

When intimately united *displacement* of one of the kidneys often occurs so that both lie on the same side of the spine, the one over the other. Complete fusion of the kidneys sometimes results in a median, flattened renal mass which usually lies in the vicinity of the pelvic brim, sometimes below the promontory of the sacrum, within the pelvis. The common sinus contains a single or double pelvis, from which proceed one or more ureters. Rarely the fused kidneys occupy a lateral instead of a median position. Displacement of the kidneys is usually associated with abnormal arrangement of the renal arteries and veins, the former springing from the adjacent portion of the aorta or common iliac arteries, the veins ending in corresponding localities into the inferior cava or common iliac veins.

Lobulation of the kidney depends upon the persistence of a condition which, while normally seen during fœtal life, ordinarily disappears in the human fœtus before birth. The superficial areas correspond to the bases of the pyramidal lobules of which the immature human kidney is composed.

The Ureter.—Doubling of the excretory canal of the kidney, on one or both sides, is occasionally observed. It may involve the renal pelvis alone, or extend to the ureter, either to the upper part or throughout its entire length. In the latter case the ureters open by independent orifices into the bladder. Gallusser[108] reported the presence of four ureters, two for each kidney, three of which opened into the bladder, while one ended blindly. The ureters may be abnormally short owing to pelvic displacement of the kidney. They are also the seat of irregular constrictions leading to congenital cyst formations.

Abnormal situations at which the ureters may terminate include the seminal vesicles, prostatic urethra and rectum in the male, and the urethra, the vagina, and uterus in the female. The explanation of these aberrant endings is found in the early embryological relations of the Wolffian duct, the cloaca, and the Müllerian duct. Since both ureter and seminal vesicle are the outgrowths from the Wolffian duct, unusual proximity of their embryonal relations may result in later union. Failure in the normal migration of the lower end of the ureter, whereby its point of termination in the urogenital sinus (later the prostatic urethra) is changed to the bladder, results in persistence of the primary close relations to the vas deferens, the representative of the Wolffian duct. Early displacements may bring the primitive ureter into abnormal relations with the hind gut, resulting in the subsequent ending of the ureter in the rectum. The early close association between the primitive renal, Wolffian, and Müllerian ducts accounts for the exceptional ending of the ureter in the vagina or uterus. Occlusions of the urinary duct in consequence of congenital narrowing and closure of the renal pelvis or ureters may take place, resulting in obstruction to the escape of the urine and distention of the tube.

The Bladder.—The urinary bladder is the seat of profound malformations resulting from arrested and imperfect development affecting the cloaca and the allantoic duct. The latter, an outgrowth from the gut tube, extends from the antero-ventral end of the cloaca for a limited distance within the belly-stalk, its intra-embryonic segment contributing the urachus and the upper part of the bladder.

While usually losing its lumen and persisting as a fibrous cord, the median vesical ligament, the urachus not infrequently remains partially pervious in its lower portion, forming either a narrow tube, lined with epithelium, or a small cyst. Exceptionally the pervious portion may communicate with the bladder and become distended. Occasionally, particularly when obstruction of the urethra takes place at an early period, the urachus may retain its lumen throughout and exist after birth as a tube which connects the bladder with the exterior of the body at the umbilicus, constituting a *uracho-vesical fistula*, through which urine may escape. In rare instances persistence of the urachus may lead to the formation of a fissure at the umbilicus of sufficient size to permit the escape of the bladder (*ectopia vesicæ*).

Vesical fissure, the most important malformation of the bladder, is associated with cleft ventral body wall, of greater or less extent, through which the posterior vesical wall appears as a red mucous surface, usually somewhat modified. The orifices of the ureters are often visible. The edges of the cleft bladder are attached to those of the external fissure, the integument and the lining of the bladder being continuous. This condition of *vesical exstrophy* may give place to complete prolapse or *inversio vesicæ*. When the cleavage is extensive it may reach from the umbilicus as far as the anus, involving the pelvis, abdominal walls, and external genital organs (*fissura abdomino-vesico-genitalis*). Vesical fissures, much more common in the male, are frequently associated with rudimentary development of the penis and dorsal cleavage of the urethra (*epispadias*). When the vesico-abdominal cleft is extensive, the posterior bladder wall is sometimes subdivided more or less completely, projecting prolapsed portions of the gut tube intervening. Pubic fissure when present may be accompanied by prolapse of the bladder and gut, causing wide separation of the halves of the cleft external genital organs, including penis or clitoris, scrotum or labia majora, as well as displacement and prevented fusion of the Müllerian ducts, resulting in double vagina and uteri.

Complete absence of the bladder, uncomplicated by other malformations, in consequence of failure of differentiation, occurs very rarely; in such cases the ureters open into the urethra, since the latter then represents the part of the cloaca which normally contributes the lower part of the bladder. Abnormal smallness of the latter is not infrequent. Subdivision of the bladder into two compartments, a larger upper and a smaller lower, sometimes occurs, the constriction in a general way indicating the extent of the allantoic and cloacal portions of the organ. Longitudinal division of the bladder, varying in completeness, has been rarely noted and probably depends upon doubling of the allantoic duct.

The attempt to explain satisfactorily the production of vesical fissure has occasioned much discussion, authorities even at present being by no means in accord. The older theories attributing the cleavage of the vesical and body wall to mechanical influences, such as overdistention of the bladder or rectum by excretions, or unusual traction of the umbilical cord, may be dismissed as entirely inadequate. Zander[109] regards persistence of the dorsal concavity, which normally for a time distinguishes the contour of the early human embryo, as an important factor

in the production of these malformations. Failure to assume the normal ventral curve has been attributed to unusual constriction or attachment of the amnion. Although still uncertain as to details, it may be assumed that these clefts are the result of a very early arrest of development involving the primary structures concerned in forming and in differentiating the cloaca, urogenital sinus, and rectum. The studies of Retterer[110] and of Keibel[111] have thrown much light upon the close relations of the cloaca to the normal development of the bladder. The last-mentioned investigator regards the fissures in question as the result of primary imperfection in the development of the ventral walls and bladder referable to faulty closure of the primitive streak. However that may prove to be, it is certain that the defects under consideration are due to a very early developmental arrest.

The Urethra.—The urinary canal, in both sexes, may be wanting, partly occluded or narrowed, due to imperfect development and more or less marked closure of the urogenital sinus. The urethra in the male consists of two embryologically distinct portions. The one, corresponding to the prostatic and membranous segments, is derived directly from the urogenital sinus; the other is formed by the prolongation and fusion of the primary genital folds along the under surface of the developing corpora cavernosa and includes the penile portion of the urethra. Faulty development and coalescence of these folds result in the production of a cleft and imperfect urethra, the abnormal termination of which may be at any point along the under surface of the more or less rudimentary penis. This condition, known as *hypospadias* and relatively common, depends upon faulty development and imperfect fusion of parts which normally exist in duplicate and unite. In the lowest grades of the defect the cleft is limited to the glans; in extreme cases the fissure may extend not only to the root of the penis, but also involve the scrotum, the component halves of which remain separate and unfused, the urethra being open as far as the prostatic segment.

Epispadias, in which the urethral cleft is associated with dorsal fissure of the penis or clitoris, is not only comparatively rare, but much less evident in its mode of production. Since the genital eminence, giving rise to the chief part of the penis or clitoris, exists as an unpaired anlage, the cleavage implies a profound defect and impression at an early period. This malformation is usually associated with vesico-abdominal fissure, very rarely existing alone, hence its genesis is usually regarded as closely identified with the influences producing the more general clefts. In many cases the dorsal groove exposing the urethra, when coincident with vesico-abdominal fissure, extends as far as the bladder, into which it opens in the vicinity of the trigonum. Doubling of the urethra sometimes occurs in consequence of faulty union of the two component halves. In a case described by Löw[112] the two canals opened by independent orifices in the navicular fossa, the smaller and upper tube extending as far back as the prostatic segment.

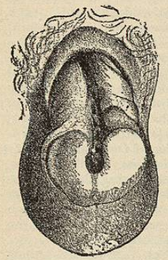

FIG. 4685.—Epispadias. (Hahn.)

MALFORMATIONS OF THE SEXUAL GLANDS AND THEIR DUCTS.—*The Testicle.*—Congenital absence of one or both male sexual glands has been rarely observed. More frequently the defect is only partial, affecting either the testicle itself or its excretory duct. In addition to the imperfections manifested in the epididymis and vas deferens in sympathy with defects of the testicle, these structures may be the seat of independent faulty development.

The most common defect of the testicle is its malposition, occasioned usually by incomplete descent into the scrotum, a migration normally accomplished shortly before birth. The undescended testicle may be retained within the abdomen (*cryptorchism*), either in its primary lumbar position or lodged at the inner end of the inguinal canal. Displacements within the abdominal wall, in front of the external abdominal ring, or within the perineum constitute respectively the inguinal, pubic, or perineal forms of ectopia. , One or both testicles may be affected, and, as a great rarity, both organs have been seen to occupy the same peritoneal sac. Frequently the retained organ suffers atrophy, or at least fails to undergo perfect development at puberty.

Malposition or even complete inversion of the descended testicle within the scrotum sometimes takes place in consequence of abnormal attachment and traction of the ligamentum scrotale.

Doubling of the testicle has been observed (Lossen[113]) as a very rare malformation which depends upon subdivision of the primary anlage of the sexual gland. The presence of a single common excretory duct in such cases is explained by the independent origin of the latter from the Wolffian body.

The Ovary.—The absence of both ovaries rarely observed, as well as the more common deficiency of one, is usually associated with other malformations of the generative organs. Sometimes, however, such defects exist along with well-formed oviducts and uterus. In most cases it is probable that absence of the ovary is due to abnormal relations and attachments of adjacent structures, in consequence of which the development of the ovarian anlage is arrested and atrophy follows.

Not infrequently the ovary remains rudimentary, its function as an egg-producing organ being never, or at best only imperfectly, assumed. The ovary may be doubled (Civatte[114]), the supernumerary organ originating from constriction and isolation of a part of the primary anlage, or of the young organ at very early stage.

Malposition of the ovary may occur in consequence of the gland becoming engaged in the inguinal canal and descending into the labium majus. At other times it escapes through the femoral or the obturator canal, or suffers displacement within the pelvis due to abnormalities of the uterus and its attachments.

The Oviducts.—Entire absence of the Fallopian tubes is usually associated with grave malformations of other portions of the generative tract; on the other hand, normal oviducts may exist when the uterus and vagina are defective. Local arrest of the development of the tubes is exhibited in the stenosis and atresia, which may affect either the uterine or abdominal orifices or the intervening portions of the duct, which are at times reduced to a solid cord. Supernumerary abdominal orifices are also encountered (Nagel[115]).

The *uterus* and the *vagina* are subject to malformations arising largely in consequence of imperfect development and faulty fusion of the two Müllerian ducts of the major part, of which these organs are the direct representatives. Normally during the third month of fœtal life the portions of the Müllerian tubes included within the genital cord unite to form a single canal, the upper part of which becomes the uterus, the lower the vagina. The fundus of the uterus for a time is divided into two horns, which are directly continuous with the oviducts, the ununited portions of the Müllerian ducts. Later the uterine horns become incorporated with the fundus.

Imperfect fusion of the Müllerian ducts throughout areas in which union normally takes place results in more or less complete doubling of the uterus and vagina. The duplicity may be effected by a septum within an externally apparently single organ, thereby producing a *bilocular* uterus or vagina. The partition is often limited to one part of the uterus, as the fundus or the cervical segment, or it may imperfectly divide the vagina.

The results of imperfect external fusion of the Müllerian ducts are seen most markedly in the uterus, which then retains the early *bicornate* condition in varying degrees. When the duplicity is restricted to a slight inden-