THE LOWER URINARY TRACT IN CHILDHOOD

KJELLBERG · ERICSSON · RUDHE

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Some Correlated Clinical and Roentgenologic Observations

BY

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PREFACE

THIS BOOK is based on clinical and roentgenologic studies of 1,155 cases of lower urinary tract disorder investigated at the Pediatric Clinic of Karolinska Sjukhuset, Stockholm, between January 1952 and April 1956. The work was carried out by a team consisting of one urologist and two roentgenologists.

It is our hope that this book will arouse increased interest in the urologic disorders of childhood, particularly with a view to early diagnosis for the prevention of severe and lasting damage. Since our main object was to give an account of our experience, the book has no pretentions to being an exhaustive survey of all the relevant problems.

All the patients at our clinic with epi- and hypospadias, as well as exstrophy of the bladder, have been treated by the plastic surgery unit, and their results will be reported elsewhere.

Space does not permit us to thank all those who have contributed in various ways to the preparation of this book. We nevertheless take this opportunity of expressing our particular gratitude to the following:

To the Staff of the Pediatric Clinic and of the Roentgenologic Research laboratory, for stimulating interest and generous help.

To Melvin Figley, for his invaluable assistance in editing the manuscript and for much constructive criticism.

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To Erica Odelberg, who translated the book from the Swedish.

Stockholm
March, 1957

THE AUTHORS

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CHAPTER I

EMBRYOLOGIC SURVEY

MANY ASPECTS of the development of the urogenital system are still veiled in obscurity, despite the enormous amount of work that has been devoted in the course of years to uncovering its problems (6, 34, 62, 85, 108, 109, 116, 120, 126, 137, 146, 147, 151, 167, 184, 185, 187, 188, 211, 212, 218, 285, 288, 289). Although the main features of the embryologic process are well understood, the details are often the subject of highly divergent opinions. In this chapter, we shall confine ourselves to an account of those details that we consider to be of importance for an understanding of the malformations occurring in our case material. Since, during fetal development, the vesico-urethral and recto-anal regions are so intimately related (9, 112), the discussion will cover certain aspects of the latter as well.

1. *Formation of the Cloaca*

The first primordium of the embryonic body has the appearance of a round disk, continuous on one side with the *amniotic cavity*, and on the other with the entoderm-clothed *yolk sac* (Fig. 1). As the *embryonic disk* increases in size, it bulges successively into the concurrently enlarging *amniotic sac*. The *amnio-ectodermal junction*, initially situated peripherally, is thereby shifted toward the ventral aspect of the disk. The growth of the fetus is most rapid in the antero-posterior dimension. As a result, the ends of the disk bulge more into the amniotic cavity than do its lateral parts (Fig. 2). Since the disk overflows its footpoint, so to speak, a circular fold develops ventrally, and penetrates most deeply in the cephalic and caudal ends, forming the *head* and *tail folds* (Figs. 1 and 2). Since the entoderm also partakes in this folding-in process, two saccular continuations, the *fore-gut* and *hind-gut*, are formed from the yolk sac, their open ends being known as the *intestinal portals* (Fig. 2). Even before this fold formation starts, an outpouching from the yolk sac into the *body stalk*, the *allantoic diverticulum*, is visible (Figs. 2 and 3). In man, this diverticulum grows for a variable distance into the body stalk, but never reaches the chorion and never becomes an allantoic vesicle.

The entoderm at each end of the primitive gut-tube lies in direct contact with the ectoderm, and fuses with it into two plates, of which the anterior is known as the *pharyngeal membrane* and the posterior as the *cloacal membrane* (Figs. 2 and 4). During the subsequent growth of the fetus, both the head and the tail part become rolled in. This displaces the cloacal membrane so that it faces cephalad and dorsally, and the intra-embryonic part of the allantoic cavity so that it runs fairly parallel to the hind-gut (Figs. 5 and 6). That part of the hind-gut lying caudally to the allantoic opening is known as the *cloaca*. It is found in its primitive state in the lower animals, in which

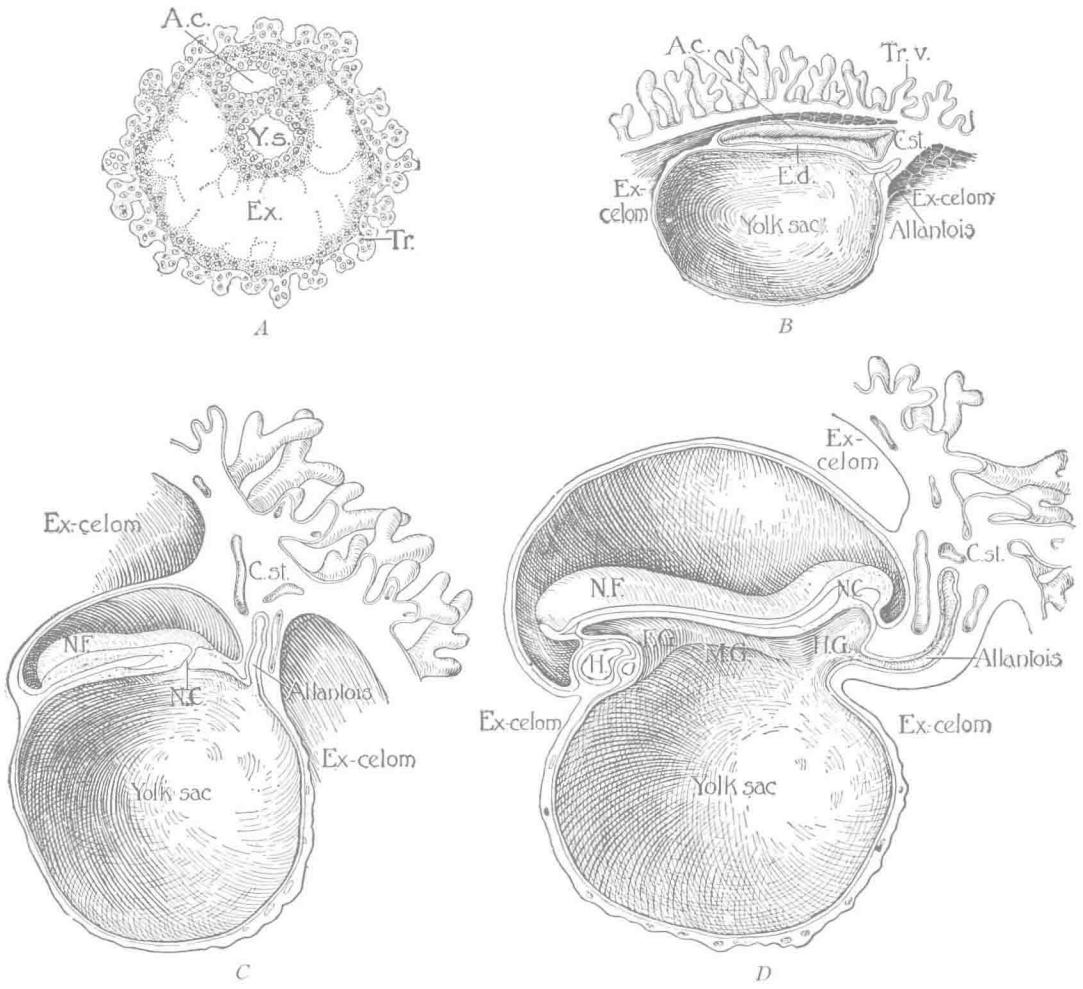

FIG. 1. Human embryos of the third week. *A*, Section; *B–D*, right halves of models. *A. c.*, Amnion cavity; *C.st.*, body stalk; *E.d.*, embryonic disc; *Ex.*, extra-embryonic coelom; *F.G.*, fore-gut; *H.*, heart; *H.G.*, hind-gut; *M.G.*, mid-gut; *N.C.*, neurenteric canal; *N.F.*, neural folds; *Tr.*, trophoblast; *Tr.V.*, chorionic villi; *Y.s.*, yolk sac. (From Arey, L. B.: *Developmental Anatomy* Philadelphia, W. B. Saunders Company 1954.)

it serves as a passage for fecal, urinary and reproductive products. With modification of the cloaca for sexual purposes, it becomes less capable of serving as a fecal passage (it becomes elongated and able to undergo eversion and intromission). In reptiles, a tendency can be noted to separation of the fecal outlet from the other passages (Fig. 7), but this separation becomes complete only in mammals (149).

2. *Division of the Cloaca*

The cavity formed by the caudal part of the hind-gut is the primordium of the *entodermal cloaca* (Figs. 5 and 6). Primarily, the tail-gut and the allantois also open into this cavity, which soon receives the two Wolffian ducts as well. As already mentioned, the ventral wall of the cloaca lies in direct contact with the ectoderm, and fuses

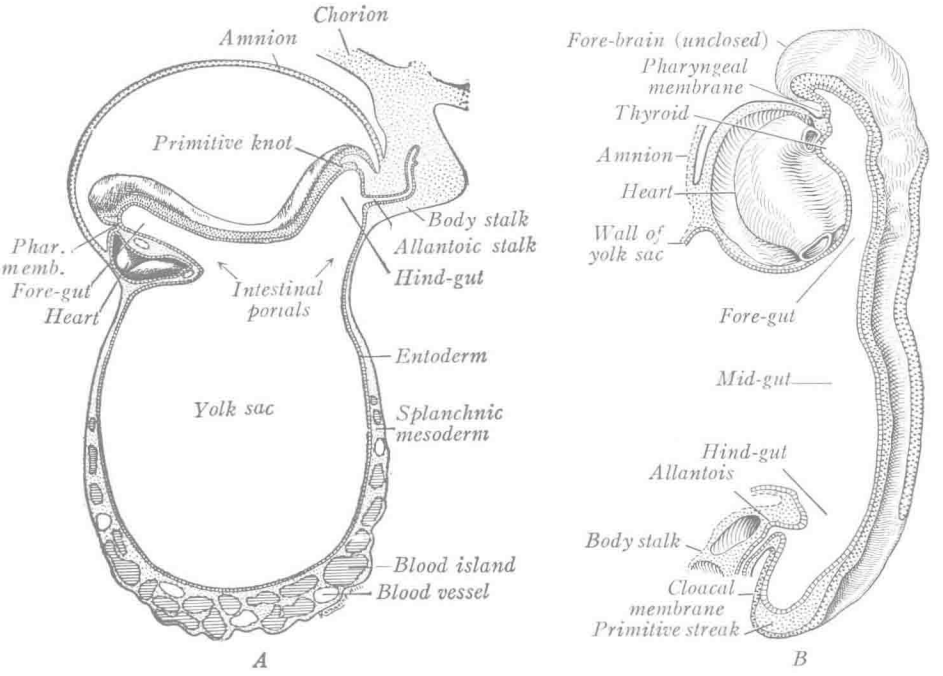

FIG. 2. Entodermal tract of early human embryos, in sagittal section. *A*, At seven somites (Prentiss, after Mall; × 23). *B*, At ten somites (after Corner; × 30). (From Arey, L. B.: *Developmental Anatomy*. Philadelphia, W. B. Saunders Company, 1954.)

with it into a single membrane, the *cloacal membrane*. This in turn forms the floor of an external, shallow indentation (Figs. 8 and 9), the *ectodermal cloaca* or *primitive proctodeum* (269). The cloacal membrane, which extends from the *primitive streak* (Figs. 4 and 6) to the base of the body stalk, is surrounded by mesenchymal tissue; this rapidly proliferates, and its ectodermal covering bulges into a *genital fold* on each side (Fig. 10).

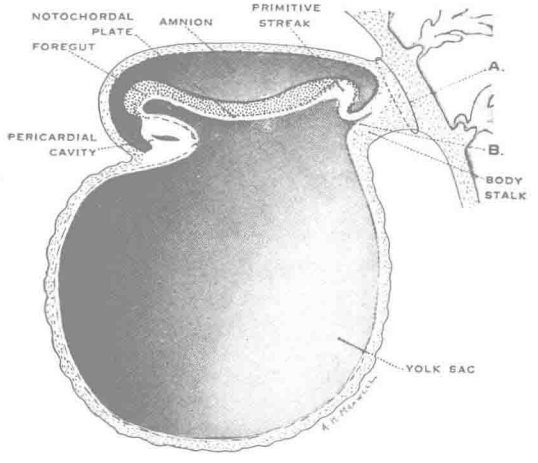

FIG. 3. Diagram to show the relations of the intrachorionic rudiment to the chorion. (From Hamilton, Boyd and Mossman: *Human Embryology*. Cambridge, Heffer and Sons Ltd., 1952.)

The postanal part of the embryo, known as the *tail bud*, grows backward and ventrally, beyond the anal plate. The hind-gut extends backward, along with the elongation of the tail bud, and forms the *postanal gut* or *tail-gut*. In a 4 mm fetus, this has the appearance of a short diverticulum, but it is rapidly prolonged as far as the tail tip (147). It is the development of the tail that brings the anal plate onto the ventral surface of the embryo. The longer the tail, the more is the postanal gut drawn out into a narrow tube, which soon atrophies and

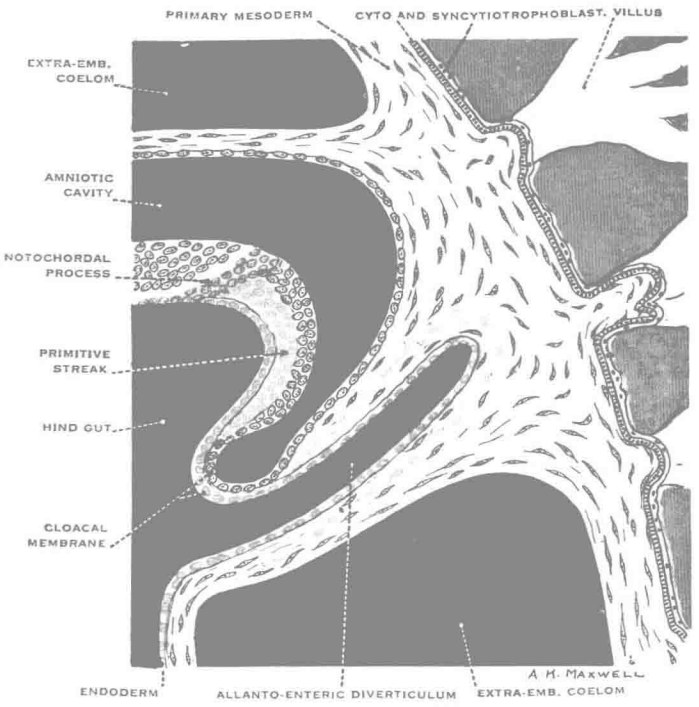

FIG. 4. The development of the tail fold and rotation of the cloacal membrane in the Sternberg embryo (3–4 somites). (From Hamilton, Boyd and Mossman: *Human Embryology*. Cambridge, Heffer and Sons Ltd., 1952.)

disappears (Fig. 5, *B* and *D*). The tail-gut is given off from the cloacal part of the hind-gut, directly beside and dorsal to the cloacal membrane; consequently, this part of the gut, if it were found in a full-term fetus, would serve as an indicator of the position of the cloacal membrane. A case is described in the literature which might have a remnant of the tail-gut (Keith (149), St. Bartholomew's Hospital, No. 3648 B). Keith stated that the specimen consists of a rectum "with a cyst attached to its posterior wall; it is exactly in the position and of the form one would expect a derivative of the post-anal gut to occupy". If this surmise is correct, the inner limiting surface of the cloacal membrane should have been at the level of the origin of this cyst in the rectal wall. Unfortunately, no description relative to this possibility is given.

Boyden (28) made a careful study of the reduction of the *hind-gut* in the ostrich. He showed that when the tail-gut disappears, excessive degeneration also involves the narrow, postero-inferior part of the cloaca, which in man would correspond to the terminal *rectum*. Ladd and Gross (160) believe that development takes place in the same way in man, and that this explains those abnormalities in which the rectum ends blindly at a considerable distance above the imperforate anus. We also share this view.

The developing *entodermal cloaca* becomes flattened from side to side and elongated in the sagittal plane. A fold, with its tip pointing caudally, is formed between the allantois and the hind-gut (Fig. 5). This fold, the *urorectal septum*, grows caudad toward the cloacal membrane and divides the cloaca, first incompletely and then completely, into a dorsal part, the *rectum*, and a ventral part, the primitive *urogenital sinus*. The latter is considered to form the common primordium of the *bladder*, *urethra* and *urogenital sinus proper* (see p. 15). In a 6 mm fetus, the

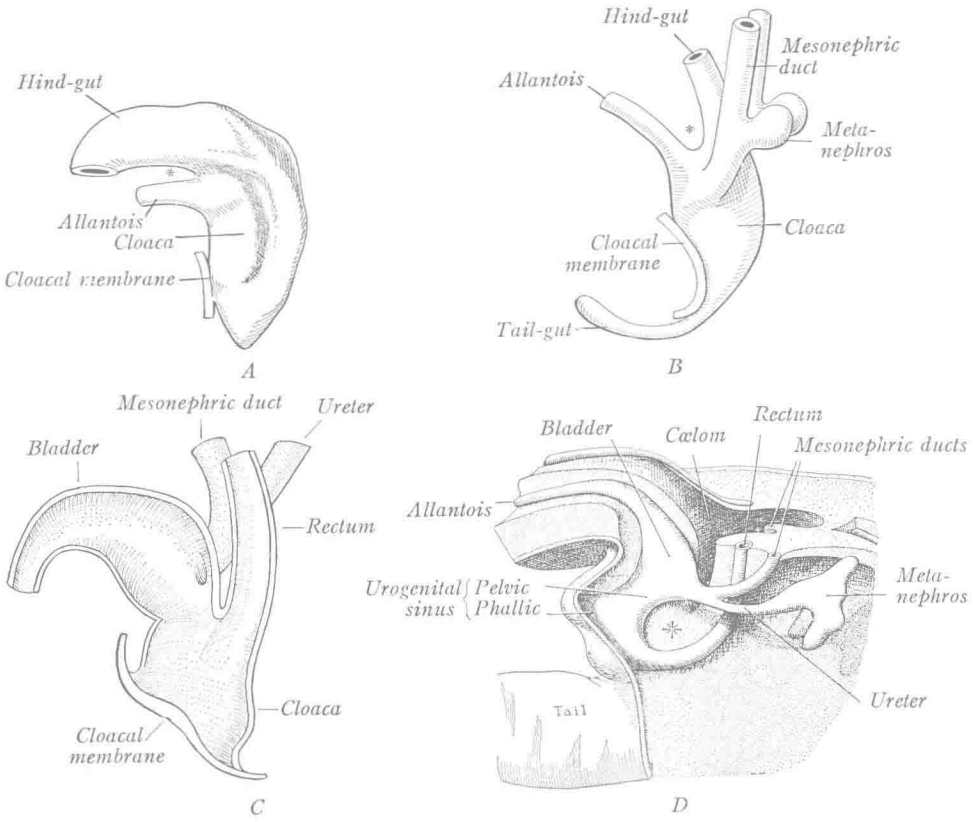

FIG. 5. Partial division of the human cloaca, illustrated by models viewed from the left side. *A*, *B*, At 3.5 mm and 4 mm, respectively (after Pohlman; × 50); *C*, at 8 mm (× 50); *D*, at 11 mm (after Keibel; × 25). An asterisk indicates the position of the cloacal septum in *A*, *B*. (From Arey, L. B.: *Developmental Anatomy*. Philadelphia, W. B. Saunders Company, 1954.)

urorectal septum reaches to the level of the opening of the *mesonephric ducts*. In a 16 mm fetus (according to Gyllensten (105) in an 8 mm fetus) separation of the cloaca is complete, and the urorectal septum fuses with the cloacal membrane. That part of the membrane lying behind the point of fusion is known as the *anal membrane*, and that part in front of it as the *urogenital membrane*. According to Spaulding (248), the latter is perforated in a 12 to 13 mm fetus; according to Keibel (146) in about a 16 mm fetus. The anal membrane, on the contrary, is not perforated until the third embryonic month, about the eighth week (17) in the 16 mm stage (217).

The site of fusion of the urorectal septum and the cloacal membrane becomes the *primitive perineum*.

Opinions are, however, divided with respect to the course of events in the division of the cloaca just described. According to Broman (34) and others, division of the entodermal cloaca starts in the fourth embryonic week. It takes place by the formation, on either side of the lateral cloacal walls, of a longitudinal fold, the *plica urorectalis*, which bulges increasingly into the cloaca. The folds gradually meet in the center, and fuse in the cranio-caudal direction into a *septum urorectale*. In the middle of the second month, the free caudal edge of the

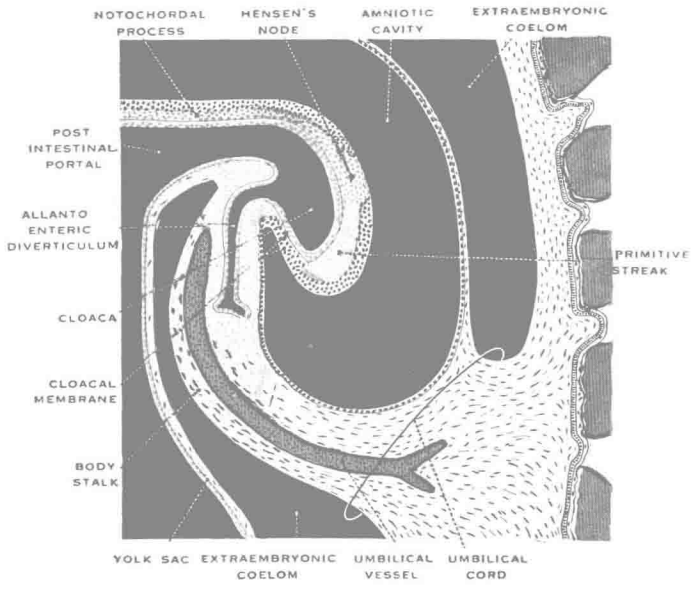

FIG. 6. Diagram to show the development of the tail fold and the rotation of the cloacal membrane in an embryo of 14 somites. (From Hamilton, Boyd and Mossman: *Human Embryology*. Cambridge, Heffer and Sons, Ltd., 1952.)

septum reaches the cloacal membrane and fuses with it.

Broman's observations are highly reminiscent of the development of the urogenital sinus in the tortoise, as described by Schmidtgen (238). He writes (p. 378): "Entstanden ist der Sinus urogenitalis durch Verwachsung von 2 Längsfalten, welche lateral von den Urogenitalpapillen entstehen". The ontogenesis thus to some extent supports Broman's view, although absence of any mark of fusion argues against his theory.

Another opinion is presented by Keith (149), who examined 114 human fetuses with malformations of the hind end of the body. He stated that the cloaca does not partake in formation of the rectum, but that the rectum is formed from the hind-gut, as its opening moves successively downward along the posterior wall of the cloaca, and finally opens separately into the proctodermal anal canal (Fig. 11) when the fetus is fully developed. Keith made a comparative anatomic study, and found that the anomalies of developmental arrest in man were directly comparable to existing developmental stages in the animal series. His views may be justified, but they do not explain every variant.

Retterer (227) also expressed the view that development of a *permanent anus* with a separate opening in man is a further advance in the process of evolution. In Monotremata (the lowest group of mammals) the rectal orifice is located, together with the genital and urinary passages, in an entodermal cloaca far from the body surface. In Marsupialia, the rectal orifice has become shifted closer to the surface, and opens into a shallow external cloaca. In Monodelphia, the process is completed, and the rectum has its own opening on the skin surface.

3. *The Mesonephric Ducts and Derivatives*

The *Wolffian ducts* (mesonephric ducts) originally open into the cloaca, close to the cloacal membrane at the 4.2 mm stage (48, 148): Figures 12 and 13. During division of the cloaca into an anterior and a posterior part, the terminal ends of these ducts are taken up into the *urogenital cloaca*. Simultaneously, the openings of these ducts

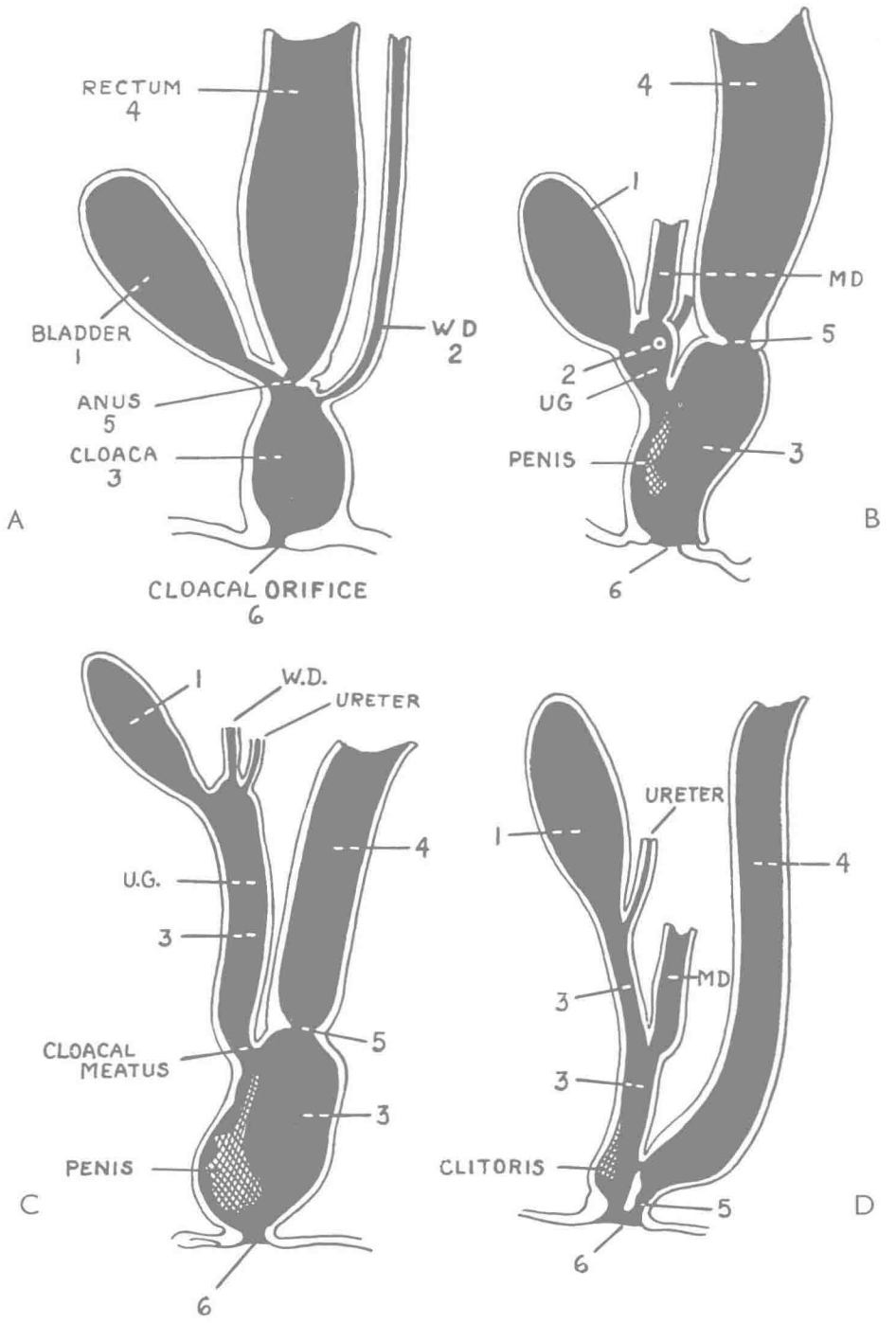

FIG. 7. Diagrams to show the manner in which the cloaca is modified and the termination of the rectum transferred from the cloaca to the perineum in higher vertebrates. (A) the amphibian form: 1. Bladder, 2. Wolffian duct (ureter and vas), 3. cloaca, 4. rectum, 5. intracloacal anus, 6. cloacal orifice. *MD* Müllerian duct. *UG* urogenital sinus. (B) form found in the tortoise, (C) form in Monotremes, (D) form found in the female marsupial. (From Keith, A.: Brit. Med. J. 2: 1736, 1908.)

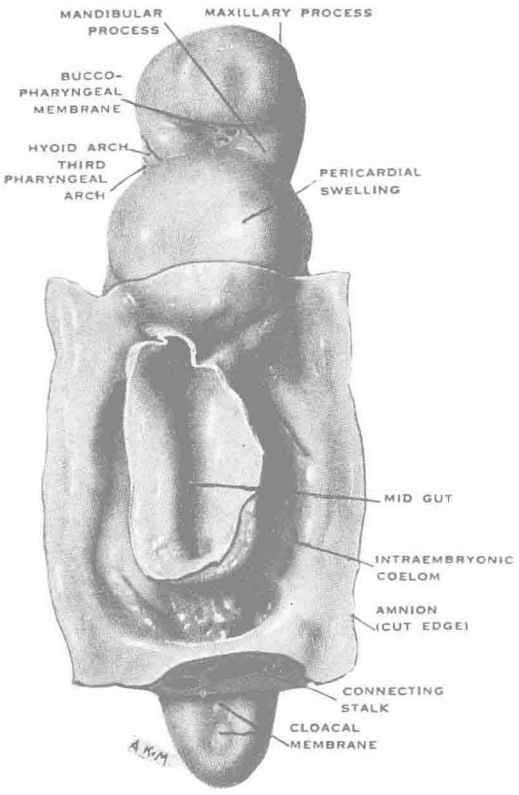

FIG. 8. The ventral aspect of a reconstruction of a 20-somite human embryo of about 36 days. (From Hamilton, Boyd and Mossman: *Human Embryology*. Cambridge, Heffer and Sons, Ltd., 1952.)

become transposed from their original site close to the cloacal membrane to the site of *Müller's tubercle*, an elevation in the future urethra. According to Stephens (253), the *inferior crest* and *fins* of the urethra (Fig. 57, p. 52) comprise the vestiges of the incorporated portions of the Wolffian ducts. The terminal fins demarcate the course taken by both Wolffian ducts in their migration from an anterior position in the cloaca to a posterior position in the urogenital sinus. When their course finally becomes more cranial, they lie side by side and form the *urethral crest*. Stephens illustrated his statements with the autopsy findings in a boy who died shortly after birth; he lacked kidneys and all the structures derived from the Wolffian and Müllerian ducts, i.e., ureters, vas deferens, seminal vesicles and ejaculatory ducts, and masculine utricle. The urethra had a normal caliber and the verumontanum was somewhat more elongated than usually. The inferior urethral crest was deficient, and no fin-like folds radiated from the submontanal region. The vesical uvula and trigone were lacking. It is known that the trigonal thickening is lacking on the side of aplasia of the kidney and ureter (11) and of single ureter with an ectopic orifice (31, 193). On the other hand, as far as we are aware, absence of the inferior crest and the fin-like folds below the

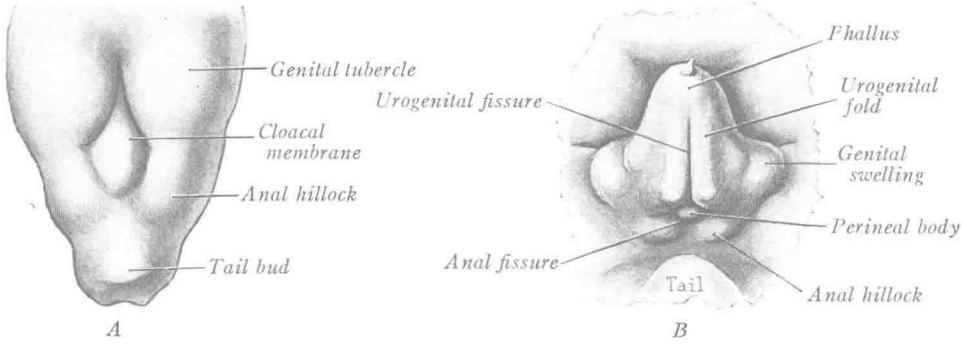

FIG. 9. Region of the human cloacal membrane, in ventral view. *A*, At 3 mm (after Keibel; ×60); *B*, at 21 mm (after Otis; ×16). (From Arey, L. B.: *Developmental Anatomy*. Philadelphia, W. B. Saunders Company, 1954.)

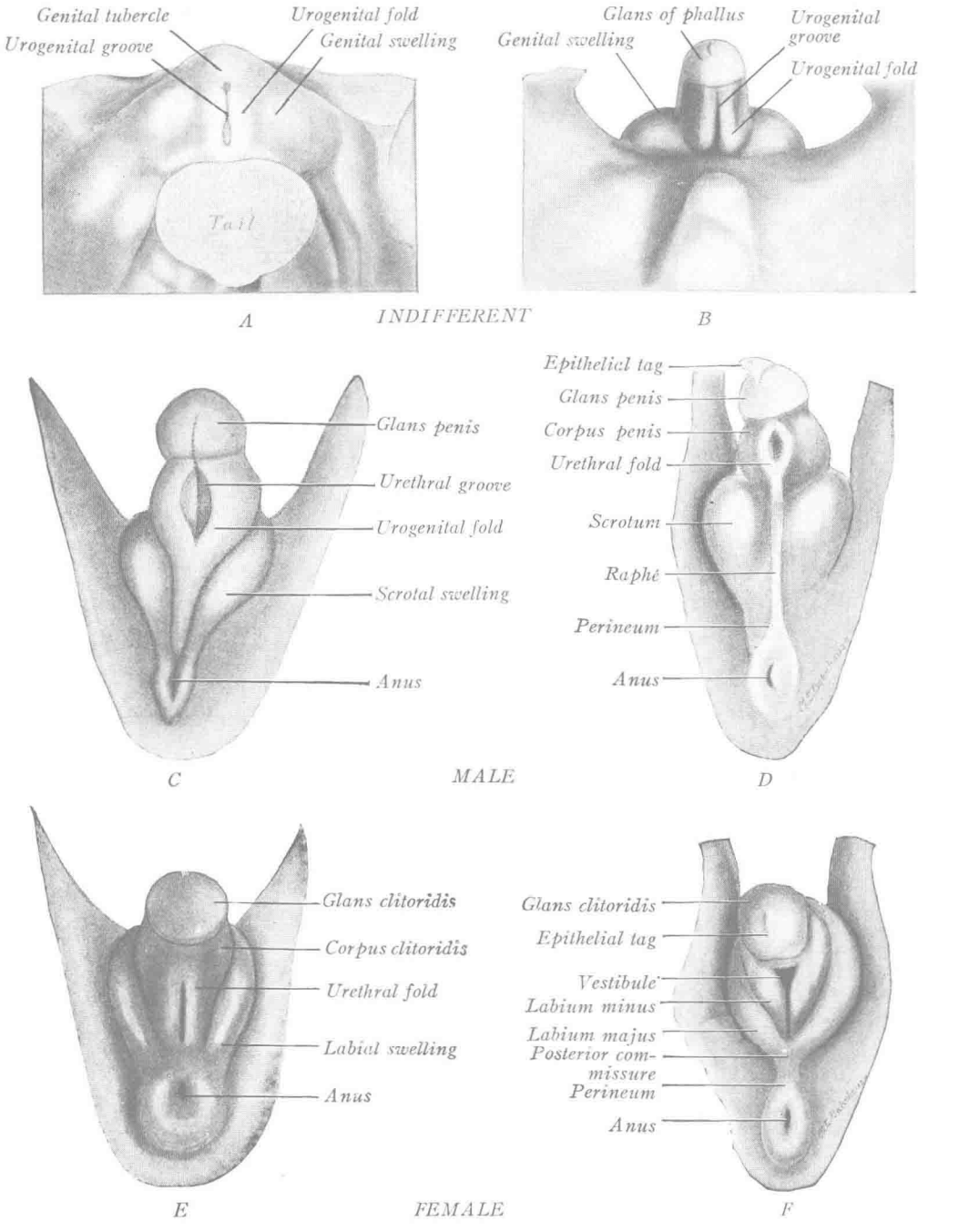

FIG. 10. Differentiation of the human external genitalia. *A*, *B*, The indifferent period, at nearly seven and nearly eight weeks. *C*, *D*, The male, at ten and twelve weeks. *E*, *F*, The female, at ten and twelve weeks. (From Arey. L. B.: *Developmental Anatomy*. Philadelphia, W. B. Saunders Company, 1954.)

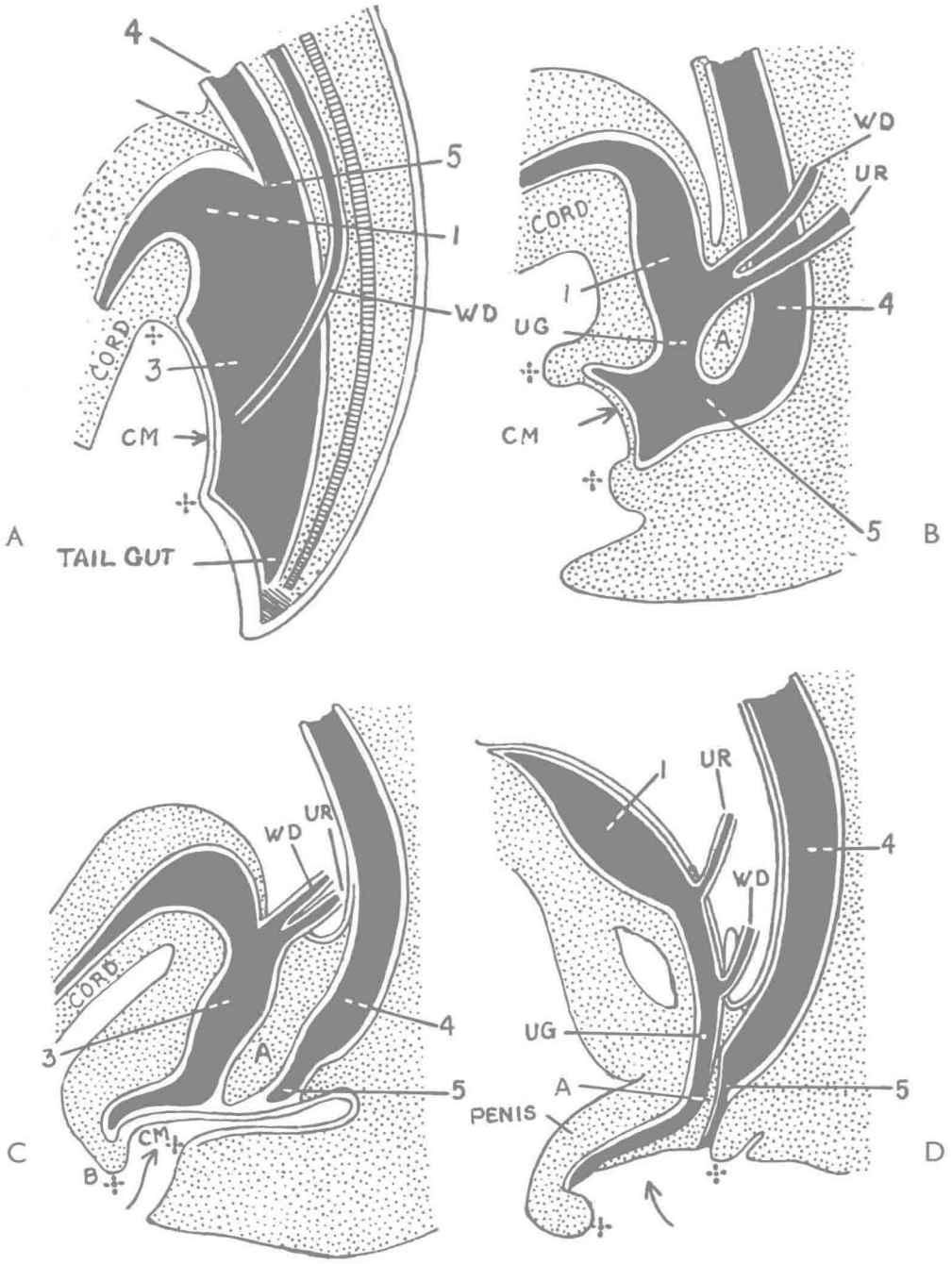

FIG. 11. Showing the manner in which the rectum becomes separated from the urogenital sinus (entodermal cloaca) during development of the human embryo. *A*, From human embryo, 4 mm long (about twenty days), after Keibel. *B*, *C* and *D*, Later stages of development: 1. Bladder, 2. Wolffian duct (ureter and vas), 3. entodermal cloaca, 4. rectum, 5. anus. *CM* cloacal membrane. *UG* urogenital sinus. *A*, mesoblast at junction of rectum and entodermal cloaca. *B*, penis, + – + the limits of the perineal depression (ectodermal cloaca). *UR*, ureter, *WD*, Wolffian duct. (After Keith, A.: Brit. Med. J. 2:1736, 1908.)