The THAI Journal of SURGERY 1998; 19:25-30.
Official Publication of the Royal College of Surgeons of Thailand

Umbilicus: Its Related Embryology, Anatomy and Pathology

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Abstract: **The umbilical cord provides the pathway for the nutrients and blood between the embryo and the chorion or placenta. Three important structures are incorporated in early fetal life within the umbilical ring and these are allantois, yolk sac and umbilical vessels. They normally lose their functions and disappear later at birth. Anomalies arising at the cord may occur as the result of the defect at the ring, patency of the allantois and urachus and present themselves with various signs and symptoms ranging from recurrent omphalitis, umbilical hernia, fistulous drainage (fecal or urinary) or even bleeding and bowel obstruction as is seen in Meckel's diverticulum. Also the summarized surgical treatments of these anomalies were reported and the basis of their management was discussed.**

The umbilicus is quite an interesting structure from its embryological, anatomical and pathological points of view. The authors' personal experience with a case of carcinoid tumor of Meckel's diverticulum and carcinoma of the urachus has prompted the review of this subject with the emphasis on the embryology and pathology.

Embryology and Anatomy[1-5]

The umbilicus occupies the central part of the anterior abdominal wall. During weeks 2-3, the extraembryonic mesenchyme condenses to form two layers. The external layer is fused to the trophoblast to form the chorion. The internal layer is attached to the amnion and with it forms the somatopleure and the yolk sac to become splanchnopleure. Between the two layers appears the extraembryonic coelom except for the area of connecting stalk where the embryo is connected to the wall of the egg. The intraembryonic coelom first appears as many small isolated spaces in the lateral mesoderm and cardiogenic mesoderm. Soon it will become the pleuro-pericardial-peritoneal cavity. The coelom divides the lateral mesoderm into two layers; a somatic (parietal) layer continuous with

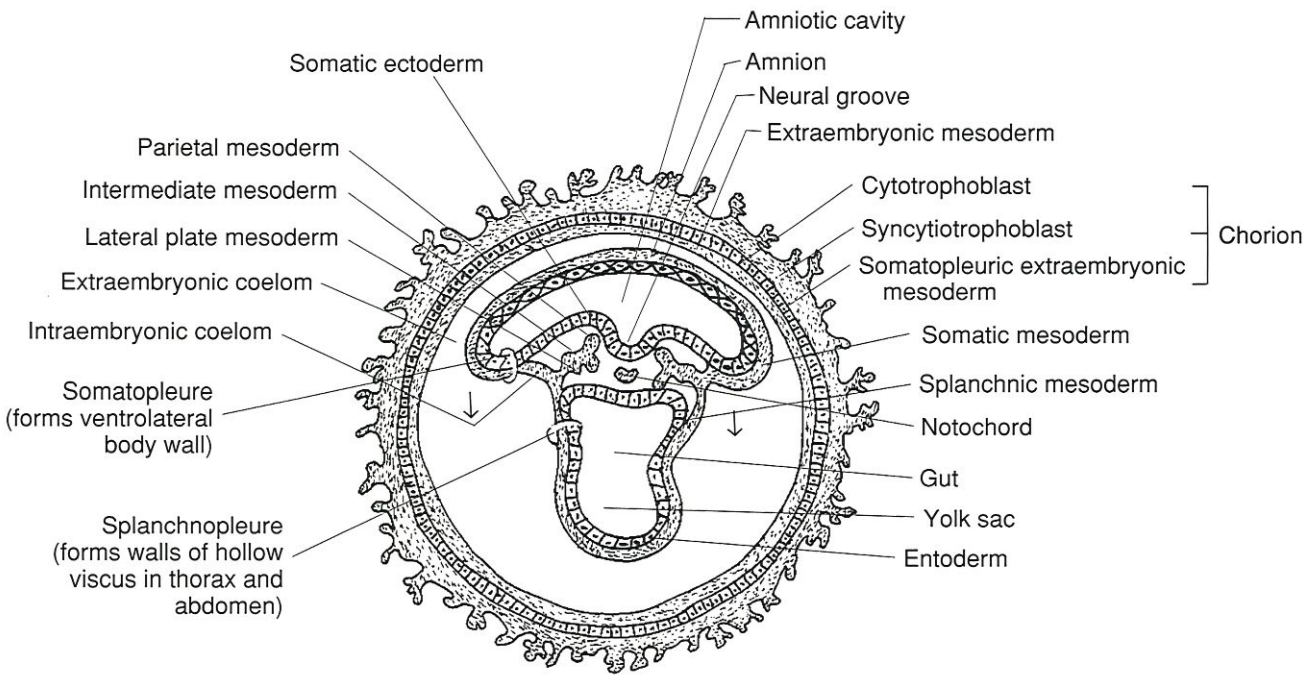

Fig. 1 Section of the embryo during second and third weeks.

the extraembryonic mesoderm over the amnion and a splanchnic (visceral) layer, which is continuous with the yolk sac. Therefore, the somatopleure forms the body wall while the splanchnopleure forms the wall of the primitive gut (Figure 1).

The extraembryonic coelom expands to form the chorionic cavity except in the area of the connecting stalk which serves to connect the embryo with trophoblast. The stalk becomes the umbilical cord with the development of blood vessels. As the amniotic cavity enlarges to encroach upon the extraembryonic coelom, the amnion, thereby forms an outer covering for the umbilical cord. The abdominal wall is formed by four separate folds-cephalic, caudal, right and left lateral folds, each of which is composed of somatic and splanchnic layers. These folds advance toward the central portion of the coelomic cavity and join at the center to form the umbilical ring. As the body wall being formed, part of the yolk sac is incorporated into the embryo as the midgut, and ultimately the connection of the midgut with the yolk sac is constricted to form a yolk stalk or vitelline duct. This narrow duct runs along the cord and connects the midgut with its distal shriveled yolk sac. The abdominal wall gradually closes off the body cavity until at birth no trace of the yolk stalk remains. The yolk stalk usually detaches from the gut by the end of week 5, but in 2 per cent of cases, the proximal intraabdominal part of the yolk stalk persists as a diverticulum of the ileum iolichis huower as Meckel's diverticulum.

The allantois appears on day 16, as a small outpouching or diverticulum from the caudal wall of yolk sac. It remains small in the human embryo and involved in early blood formation and related to the development of the urinary bladder. Blood formation occurs in its walls during week 3-5 and its blood vessels become the umbilical artery and vein. The allantois progresses in the connecting stalk along these umbilical-allantoic vessels. At week 8, the allantois after being extended over almost the entire length of the umbilical cord disappears distally while the vessels continue to develop. Therefore the intraembryonic portion of allantois connects the umbilicus to the urinary bladder. As the bladder enlarges, the allantois involutes to

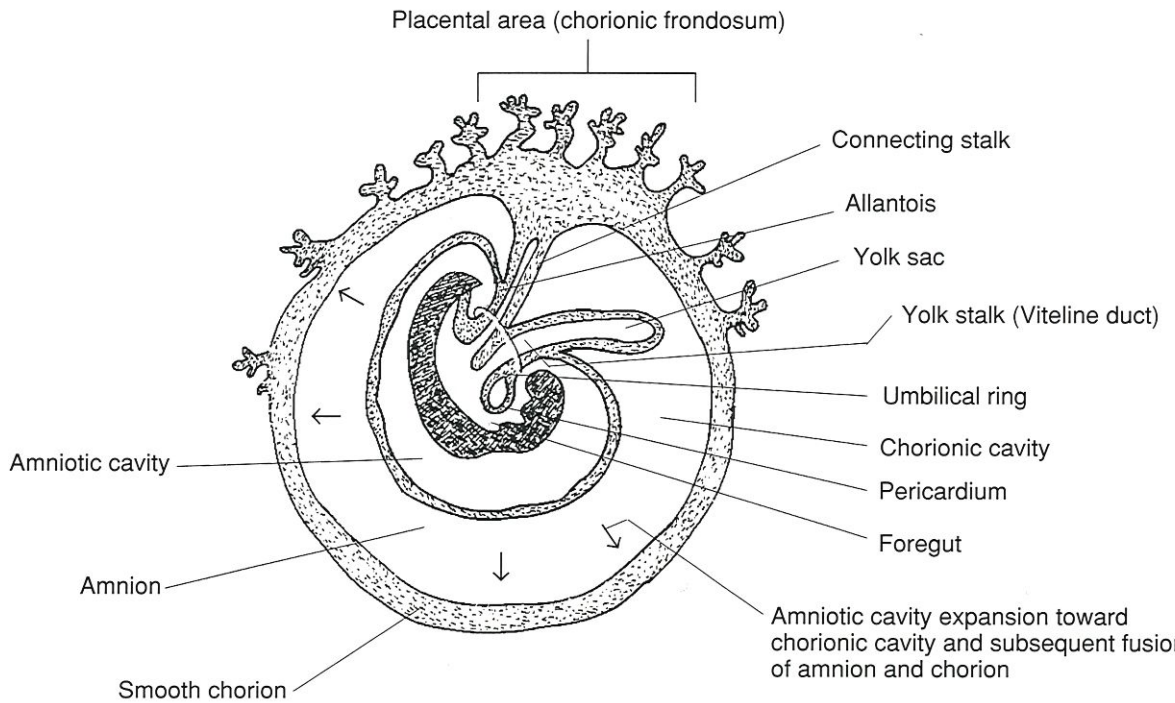

Fig. 2 Section of the embryo showing formation of allantios, yolk stalk and umbilical ring.

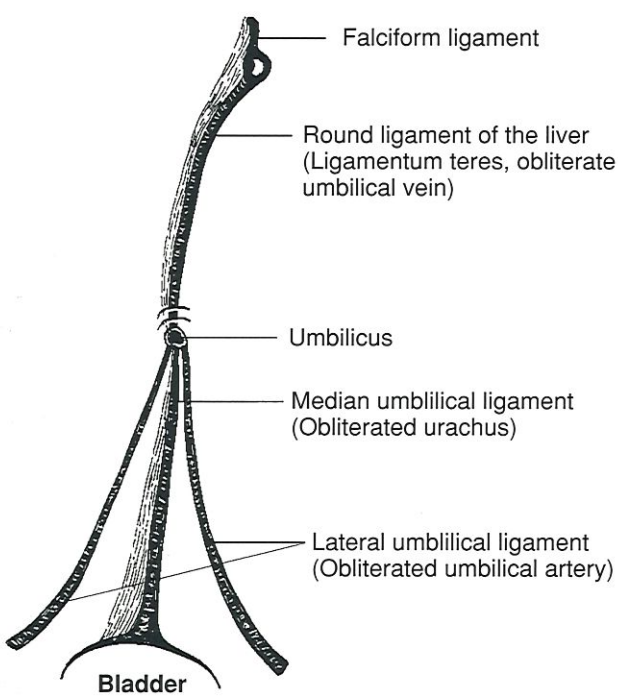

Fig. 3 Umbilical region displayed from within.

form the urachus (Figure 2).

In short, around the end of the third month, the abdominal wall has closed off except at the umbilical ring. The amnion invests the cord externally and is continuous with the skin of the abdomen. The umbilical cord encloses the following components; the yolk stalk, allantois, and the fetal blood vessels connecting with the placenta. If the peritoneal surface of the umbilical region is examined, four fibrous cords will be seen radiating from it. These are remains of four tubes which pass through the umbilical cord in fetal life; these are the urachus, the right and left umbilical arteries and the left umbilical vein (Figure 3). Originally, there were two arteries and two veins. The left vein is larger and persists but the right which is smaller later disappears before the embryo is 10 mm. long. The left umbilical vein passes upward from the umbilicus to the liver, occupying the free border of the falciform ligament but obliterated later to become the round ligament of the liver or ligamentum teres. The umbilical arteries run on either side of the urachus from the internal iliac artery to the umbilicus, when obliterated they are known as the lateral umbilical ligaments. The urachus also closes off to become

median umbilical ligament. In adult, the umbilical arteries may remain patent and form the superior vesical branches to the urinary bladder.

Pathology

Umbilical Hernia

Initially, between the fifth and tenth weeks of fetal development, the alimentary canal increases in length much more rapidly than does the coelomic or body cavity. Accordingly much of the gut extrudes itself through the large patent defect in the abdominal wall and lies within the proximal portion of the umbilical cord. As the body cavity enlarges to the size big enough to accommodate the abdominal vicera, the intestinal loops return to the body cavty. Contraction of the umbilical ring completes the process of the abdominal wall formation. Failure of the closure of the umbilical ring leaves a central defect in the linea alba. The resulting umbilical hernia is thereby covered by umbilical skin and subcutaneous tissue and the defect in the fascia allows the protrusion of the abdominal content. Usually, the hernia of less than a centimeter or one finger-tip at birth will close spontaneously by three years of age. The larger defect may require surgical repair.

Omphalocele

Large defect at the umbilical ring covered by the translucent membrane is the manifestation of the anomaly at the umbilicus which allows the protrusion of the mass consisting of bowel and solid viscera. The size may vary from about one centimeter in diameter to the huge defect containing much or all of the abdominal viscera. Allen and colleagues[4] suggested the use of Silastic material sutured around the circumference of the defect as an exterior covering without skin. This procedure provides temporary coverage of large omphalocele as well as exposed bowel in gastroschisis. The principle of this method is to provide the covering and steady pressure by gradual reduction of the plastic coverage over the interval of 5-14 days until the abdominal wall closure can be performed and tolerated by the infant.

Gastroschisis

This pathological condition, orginally thought to be a variant of omphalocele, is probably results from a fetal accident in the form of intrauterine rupture of a hernia of the umbilical cord. Shaw[6] first suggested that the gastroschisis is simply a hernia of the umbilical cord that ruptures after the complete development of the abdominal wall. This is due to the finding that the baby with gastroschisis has a lot of intestines on the surface of the abdominal wall and the defect is partly detached allowing free communication with the abdominal cavty. The intestine found outside may be glistening, moist and normal in appearance suggesting that the rupture may have occurred immediately before or during delivery of the infant.

Meckel's Diverticulum

This is the most common congenital anomaly of the gastrointestinal tract, occurring in 1-3 per cent of the population.[7] It is a remnant of the vitelline duct or omphalomesenteric duct which connects the primitive gut to the yolk sac in early fetal life. Normally, this structure is obliterated by the 7th to 8th week of gastation when the placenta replaces the yolk sac as the source of nutrient for the fetus. Failure of obliteration may result in an omphalomesenteric fistula, an enterocyst, a fibrous band connecting the small intestine to the umbilicus and finally Meckel's diverticulum. This is a true diverticulum because it arises from the antimesenteric border of the ileum and has all layers of the intestinal wall. Meckel's diverticulum with or without a fibrous connection to the umbilicus accounts for 90 per cent of all omphalomesenteric or vitelline duct anomalies. Many of them contain heterotopic mucosa of which 60 per cent are gastric mucosa. The total lifetime complication rate is about 4 per cent and decreases to near zero with old age. Over 40 per cent of those having complication are under 10 years of age. Therefore, surgical removal of an asymptomatic Meckel's diverticulum found incidentally at laparotomy in adults is not indicated because the morbidity from incidental removal (reported to be as high as 12%) far exceeds the potential for prevention of the complication.[8]

The symptomatic complications of Meckel's diverticulum fall into 3 main categories; bleeding, obstruction and inflammation. Bleeding is the most common presentation in children, usually presents as acute, episodic, painless, dark red, maroon, currant-jelly stools while melena is more likely in adults. Ob-

struction is the most common presentation in adults. Obstruction may be caused by entanglement of the small bowel around the fibrous cord, entrapment of an intestinal loop within a mesodiverticular bland (the fibrous cord formed from the omphalomesenteric vessels), volvulus, intussusception and incarceration within an adjacent hernia sac.

Because of high incidence of heterotopic gastric mucosa is present in the diverticulum, therefore, radioisotope[9] scanning using $^{99m}T_c$ pertechnetate is quite helpful to make the diagnosis. At times, angiogram[10] is used to evaluate unexplained GI bleeding since superior mesenteric artery injection may demonstrate a branch of the ileocolic artery or the remnant of right vitelline artery. Tumors of this diverticulum are rare. Up to 1959, 15 cases[11] of leiomyosarcoma had been reported. Carcinoid tumors are probably the most common neoplasms of Meckel's diverticulum. Singhabhandhu[12] B et al reported the 50th case and reviewed the literature up to 1973. This is a case of a 60 years old male which was operated with the initial diagnosis of acute appendicitis. The appendix was normal but an inflammed Meckel's diverticulum was found and so resected with short segment of smale bowel. Here, the carcinoid tumor was incidentally found.

Patent Urachus

During the development of coelomic cavity, there is a free communication between the urinary bladder and the abdominal wall through the urachus which exists side by side with the omphalomesenteric or vitelline duct. The upper part of allantois is continued into the umbilicus and the umbilical cord in early fetal life. This intraabdominal portion of the allantois is called urachus. The urachus and the allantois will normally be obliterated to become a solid cord, so called median umbilical ligament. Persistence of this tract, however, will result in a communication between the bladder and the umbilicus. The lumen in the lower urachus may persist and continuous with the bladder cavity as is seen in many cases. Other malformations include urachal sinus or dilatation of the upper urachus opening at the umbilicus. When the entire urachus remains patent, the urachal fistula will occur. The first sign of a patent urachus is moisture or urine flow from the umbilicus as a result of so-called umbilical urinary fistula. This is comparable to umbilical fecal fistula as is seen in patent omphlomesenteric duct. Malignant neoplasms[13] may arise in the urachus. The urachus often has islands of glandular epithelium and microscopically there is proliferation in this adenoma-like structures. This glandular epithelium is thought to have originated from embryologic cloacal inclusions or enteric rests or as a result from metaplasia. Singhabhandhu[14] B and Punyaprasiddhi T reported a case of mucin producing carcinoma of the urachus in a 24 years old male and reviewed the clinical management. The primary complaints of the patient were lower abdominal pain and midline lower abdominal mass for two weeks. At operation, the firm mass was found in the area extending from the umbilicus to the dome of the bladder. This mass along with the adjacent abdominal wall was excised. There were multiple metastatic nodules in the omentum and parietal peritoneum. The patient succumbed to the disease two weeks after operation from wide-spread intraabdominal metastases. The origin of the carcinoma is thought to be from totipotential primitive cloacal cells lining the urachal lumen.

References

1. Anson BJ, Maddock WG. Gallander's surgical anatomy. 3rd ed. Philadelphia: WB Saunders, 1952:375-82.
2. Thorek P. Anatomy in surgery. 2nd ed. Philadelphia, Montreal: JB Lippincott Co, 1962:373-7.
3. Pansky B. Review of medical embryology. New York: Macmillan Publishing Co, 1982:86-93.
4. Guzzetta PC, Anderson KD, Altman P, Newman KD, et al. Pediatric surgery. In: Schwartz SI, Shires GT, Spencer FC, eds. Principles of surgery. 6th ed. New York: McGraw-Hill, 1994:1706-10.
5. Gray SW, Skandalakis JE. Embryology for surgeons. Philadelphia: WB Saunders, 1972:519-27.
6. Shaw A. The myth of gastroschisis. J Pediatr Surg 1975; 10:235.
7. Turgeon DK, Barnett JL. Meckel's diverticulum. Am J Gastroenterol 1990; 85:770-87.
8. Soltero MJ, Bill AH. The natural history of Meckel's diverticulum and its relation to incidental removal. Am J Surg 1976; 132:168-73.
9. Harden RMG, Alexander WD, Kennedy I. Isotope uptake and scanning of stomach is man with 99mTc-per-technetate. Lancet 1967; 1:1305-7.

10. Klein HJ, Alfidi RJ, Meaney TF, et al. Angiography in the diagnosis of chronic gastrointestinal bleeding. Radiology 1971; 98:83-91.
11. Skandalakis JE, Gray SW, Shepard D, Bourne GH. Smooth muscle tumors of the alimentary tract. Springfield: Charles C Thomas, 1962.
12. Singhabhandhu B, Gray SW, Krieger H, Gertman KE, Skandalakis JE. Carcinoid tumor of Meckel's divertculum; report of a case and review of literature. J Med Assoc Georgia 1973; 62:84-9.
13. Perumalswamy R, Novin N. Urachal carcinoma: case report and review of literature. MD State Med J 1983; 32:770-2.
14. Singhabhandhu B, Punyaprasiddhi T. Carcinoma of the urachus. J Int Coll Surg Thai 1988; 36:41-4.