Journal of Korean Medical Science
Vol. 6, No. 2, 177-181 June 1991

# Body Stalk Anomaly
## —A Case Report—

Soon Ae Jun, M.D., Myong Ock Ahn, M.D.,
Seung Sook Lee, M.D.*, Je G. Chi, M.D.* Kyung Sub Cha, M.D.

*Department of Radiology and Department of Obstetrics and Gynecology, Cha Women's Hospital of Seoul, Korea*
*and Department of Pathology*, Seoul National University, Seoul, Korea*

***A case is presented of an amnionic rupture sequence which led to massive fetal ventral herniation and lordoscoliosis. Characteristic ultrasonographic findings of an omphalocele, fetal attachment to the placenta, and the absence of free-floating umbilical cord were observed.***

**Key Words :** *Early amnionic rupture, Body stalk anomaly, Ultrasound, Congenital anomaly*

## INTRODUCTION

Early amnionic rupture occurring between 5th and 7th gestational weeks leads to spontaneous abortion or maldevelopment of the fetus. The most severe malformation of early amnionic rupture is body stalk anomaly, which is lethal. We present a case of body stalk anomaly with antenatal ultrasonographic findings.

## CASE REPORT

A 29-year-old, gravida 2, para 0, abortion 1, pregnant woman presented to the hospital with an intrauterine pregnancy of 11 weeks. Ultrasonographic evaluation revealed a single fetus with 11 weeks. An abnormal 1.7×1cm sized mass, ventral to the fetus, was attached to the placental tissue. One part of the placental margin was elevated in a sharp peak (Fig. 1). The amount of amniotic fluid was normal. The ventral mass showed homogeneous soft tissue echo density, and the remaining fetal ultrasonographic findings were not remarkable.

At 20 weeks gestation, a follow-up ultrasonogram showed a living, breech-presented fetus with a normal amount of amniotic fluid. The biparietal diameter was 5.0cm, which was consistent with 20 weeks gestation. A severe spine deformity was noted. While the thoracic cavity looked small, the heart was normal in size and position, the movement of fetal extremities was active; however, there was no truncal movement due to an omphalocele fixed to the placenta (Fig. 2). A fetal ventral wall mass of approximately 5×6cm was noted. The mass was enclosed by an irregular thickened membrane, part of which was made up of two layers. This membrane was connected to the placenta along a broad base, and, therefore, the placenta appeared to be abruptly amputated the mass (Fig. 3). Within the sac, the liver,small and large bowels, and stomach were noticed. A 2.0×1.5cm sized nodular knot between the center of the liver and the placenta site was found. Moreover, there was a homogeneous echo-free area in the sac. The placenta was located posteriorly, and the dorsal side of the fetus faced the placenta which showed a normal texture. Free-floating loops of the umbilical cord could not be seen in the amniotic fluid.

A poor prognosis of the anomalous fetus

**Address correspondence to :** *Dr. Soon Ae Jun, Department of Radiology, Cha Women's Hospital of Seoul, 650-9 Yeok Sam Dong, Kang Nam Gu, Seoul (Tel ; (02) 557-2611)*

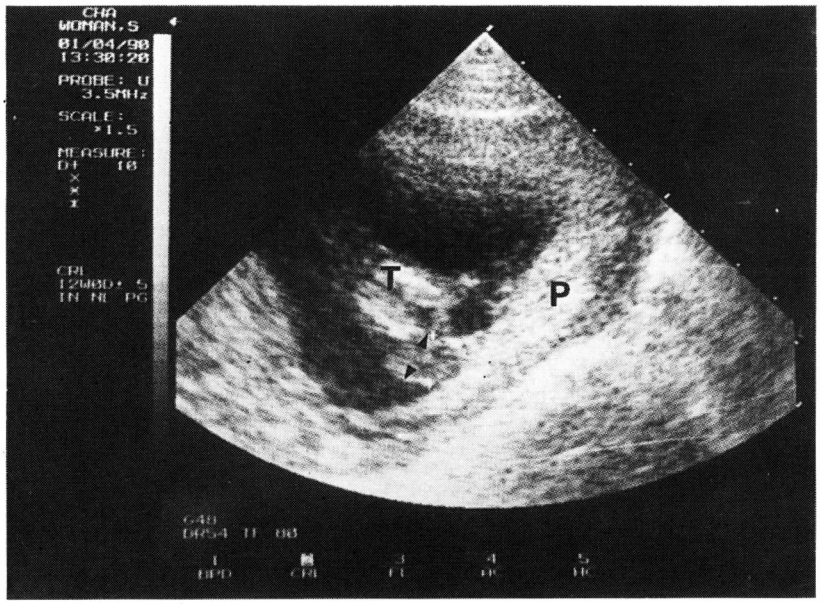


**Fig 1.** IUP 11 gestational weeks. A 1.7×1 cm sized mass, ventral to the fetus, is attached to the placenta which looks like a sharp peak. P=placenta ; T=fetal trunk ; arrowheads=abnormal mass

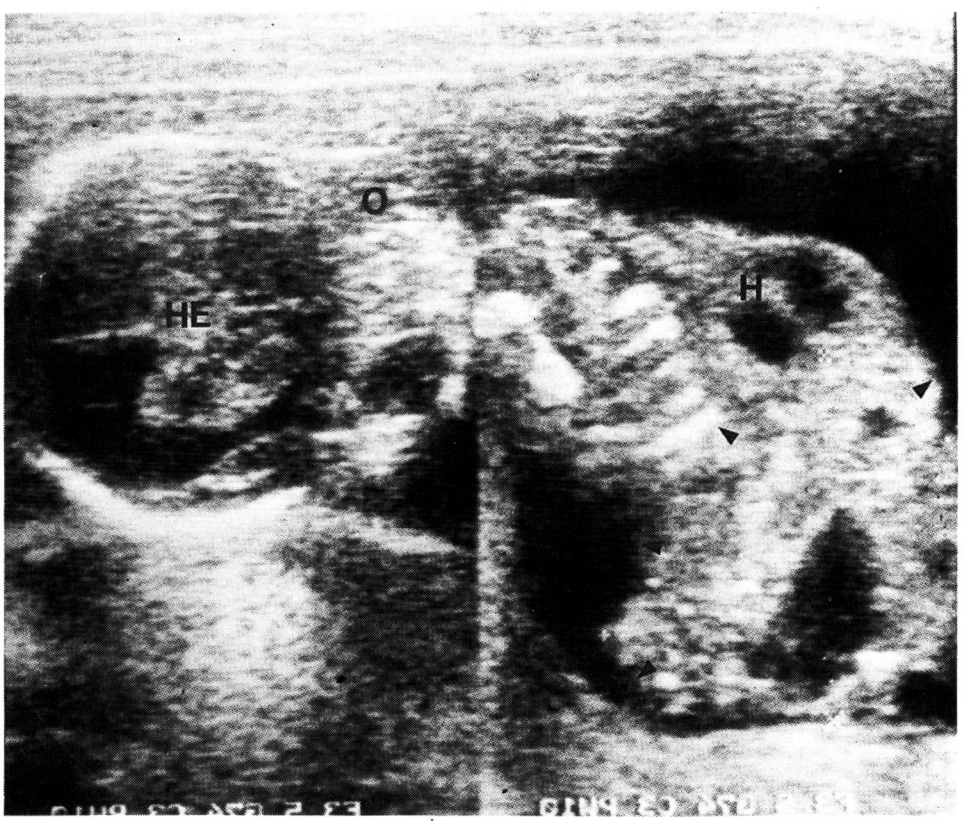


**Fig 2.** Coronal section of the fetus. IUP at 20 weeks, an abnormal 5×6 cm sized mass with a covering membrane containing the bowels. HE=head ; O=orbit ; H=heart ; arrow heads=abnormal mass

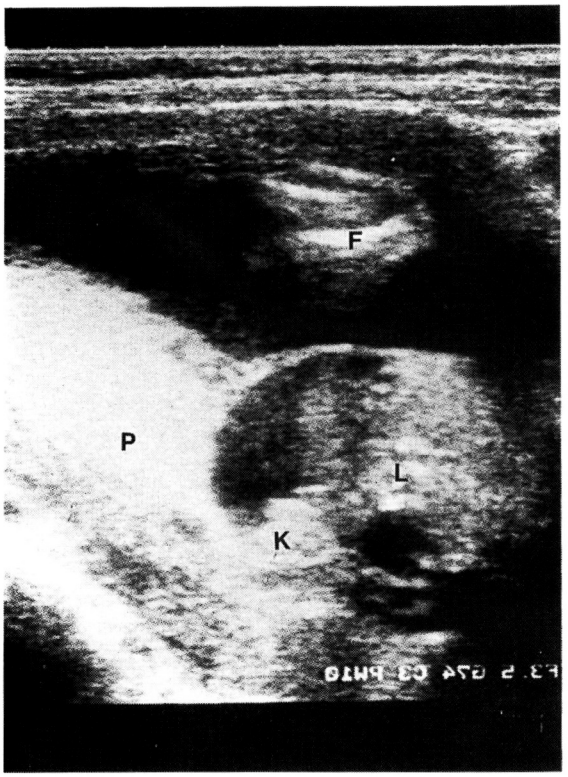


**Fig 3.** Within the sac, a 2×1.5 cm sized vascular knot, which connects the liver and the placenta can be seen. The sac, with an irregular thickened membrane, is connected to the placenta along the broad base. L= liver; K=vascular knot; P=placenta; F= femur

**Fig 4.** An external view of the fetus shows a midline defect and herniated viscera which are directly continuous with the placenta (right). Note also lordoscoliosis of the body.

was suggested, and induction of labor was tried. The patient delivered a living male infant, weighing 580 gm and the attached placenta simultaneously. The infant died immediately after birth. Chromosomal analysis of fetal cord blood sample showed a 46, XY karyotype.

Postmortem examination of the fetus and placenta confirmed the findings seen in ultrasonography (Fig. 4). Virtually no umbilical cord was identified but only umbilical vessels with a hint of Wharton's jelly being traced along the margin of the stump (Fig. 5). The placenta was attached to the herniated viscera that was covered by a thin transparent membrane. This membrane was directly continuous with the amniotic membrane of the placenta without sharp demarcation. Because of this fetoplacental attachment, the body axis became modified to show an irregular curving (lordoscoliosis). The liver, small and large bowels, spleen and stomach were included in the sac. The peritoneal cavity was narrow and contained a spherical left kidney with a double ureters, the rectosigmoid colon, pancreas, adrenal glands, and urinary bladder. The chest organs showed hypoplastic lungs (a trilobed right lung and a one-lobed left lung) and the heart. The spleen was deeply fissured in the middle and was almost separated into two parts. An accessory spleen was noted in the hilus portion. The liver was quadriangular with right-left symmetry. The thyroid showed a large isthmus. The brain was unremarkable. The placenta was discoid and weighed 150 gm. No abnormal staining or deposition was seen. A cut section showed no abnormalities. Microscopic examination of the viscera showed no recognizable abnormalities.

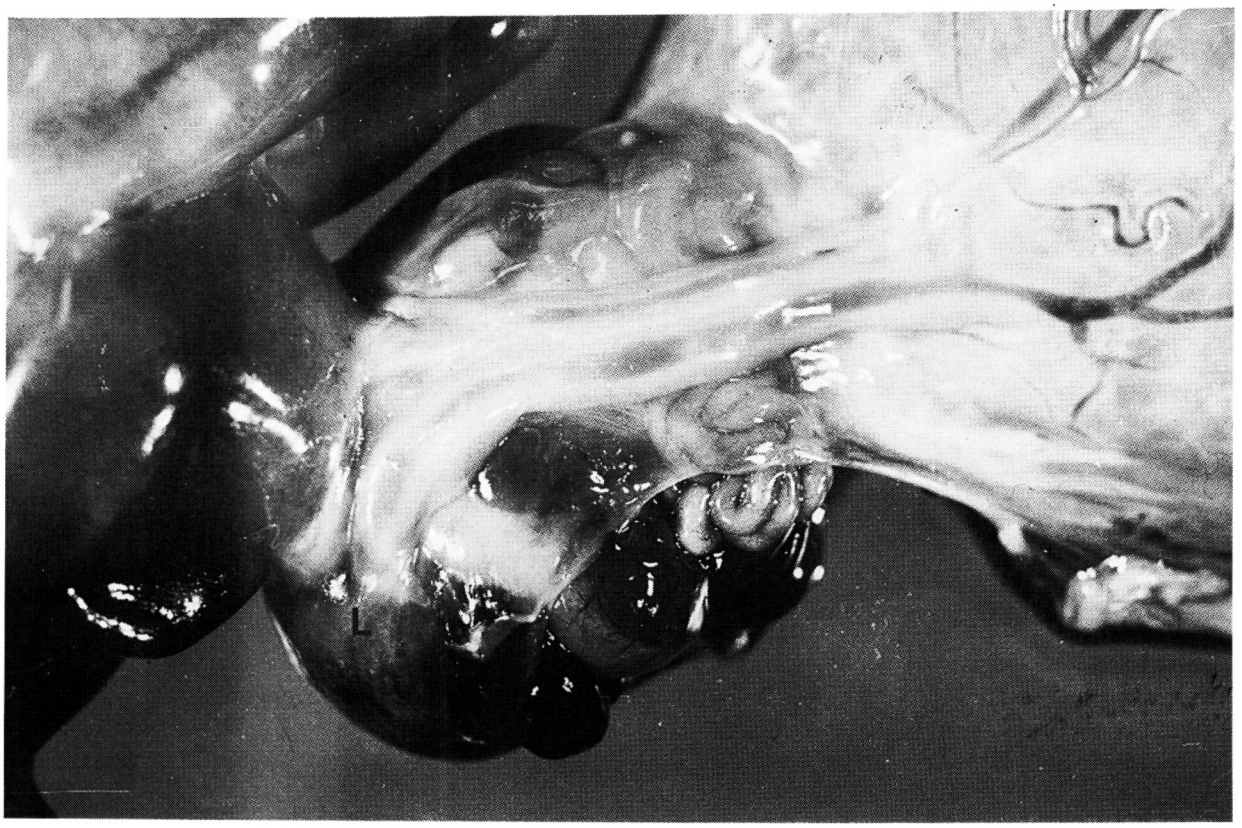


**Fig 5.** A close-up view of the body stalk anomaly. The amniotic membrane covers the herniated liver and intestinal loops. The umbilical cord is not present and is only represented by vessels that connet the chorionic vessels to the fetal liver (L) without forming Wharton's jelly.

## COMMENT

The etiology of body stalk anomaly has been regarded as an idiopathic and sporadic event, and the recurrence rate is negligible. Body stalk anomaly has been thought to be the failure of formation of the body stalk, and absence of the umbilicus and umbilical cord is the salient finding.

At the 5th gestational week, the flat embryonic disk is transformed into a cylindrical form by cephalic, lateral, and caudal foldings. By this folding process, the intraembryonic coelom (peritoneal cavity) becomes separate from the extraembryonic coelom (chorionic cavity). The failure of complete obliteration of the extraembryonic coelom accounts for the absence of umbilical cord formation so that the wide based insertion of the amnio-peritoneal membrane onto the placental chorionic plate could occur. Thus, all the organs of the intra-abdominal cavity protrude into extraembryonic coelom. The sac is made of amnion and peritoneal membrane and is attached directly to the placenta(Lockwood et al., 1986).

Body stalk anomaly is not a specific separate diagnosis but is one part of a broad spectrum of defects due to early amniotic rupture sequence. Early amnion rupture and its consequences are not rare and cause spontaneous abortions with structural abnormalities, stillbirths or early neonatal deaths. The nature and severity of the consequences of early amnion rupture relate to the timing of the event.

Early amnion rupture causes problems in morphogenesis because of early compression of the embryo, such as abnormal pla-

centa attachment to the head or abdomen, scoliosis, craniofacial anomalies, anencephaly, limb defects, abdominal wall defects, and thoracic wall defects. A late occurrence of amnion rupture only results in external surface abnormalities of the fetus with rare internal anomalies(Jones, 1988). Skeletal defects such as kyphoscoliosis or lordoscoliosis could occur because of the fixed fetal position.

Common associated anomalies are as follows : neural tube defects, intestinal atresia, thoracic wall defects, pericardial defects, heart anomaly, and single umbilical artery none of which were seen in this case ; however other anomalies, such as lordoscoliosis, pulmonary hypoplasia, liver symmetry, spherical left kidney, and double ureter were noted. The disease is uncorrectable because all of the abdominal contents protrude outside of the abdominal cavity, so the size of the abdominal cavity is too small to include all the organs in it by operation.

Sonographic features of this anomaly are fairly characteristic in fetal attachment to the placenta, omphalocele, and absence of free-floating loops of the umbilical cord in the amnionic cavity. These sets of ultrasonographic abnormalities appear to be virtually diagnostic of body stalk anomaly, which should be distinguished from conventional omphalocele, gastroschisis and other midline defect syndromes.

## REFERENCES

Lockwood CJ, Scioscia AL, Hobbins JC : *Congenital absence of the umbilical cord resulting from maldevelopment of embryonic body folding. Am J Obstet Gynecol 1986 ; 155 : 1049-1051.*

Jones KL : *Smith's Recognizable Patterns of Human Malformation, 4th ed. Philadelphia, Saunders. 1988 : 576-577.*