**Intraventricular fetus-in-fetu**

**Case report**

**FARHAD AFSHAR, B.SC., M.D., F.R.C.S., THOMAS T. KING, F.R.C.S., AND COLIN L. BERRY, M.D., PH.D., F.R.C. PATH.**

Departments of Neurosurgery and Pathology, The London Hospital, Whitechapel, London, England

The authors present a case of an intraventricular 14-cm fetus in a 6-week-old infant who presented with enlarging head size. The entire intracranial fetus, with recognizable limbs, trunk, head, and exomphalos, was removed totally from the lateral and third ventricles, with excellent recovery in the patient, who is now 18 months old. The cytogenetical and tissue histological studies are reported. The embryology, operative procedure, and pathology of this condition are discussed and the literature briefly reviewed.

**KEY WORDS**  
- fetus-in-fetu  
- incomplete twinning  
- intraventricular tumor  
- heteropagus

**FETUS-IN-FETU** is an extremely rare condition and represents a form of incomplete twinning. In some reports and texts it is confused with teratoma, despite clearly different origins and probable development. Willis stated that “included twins have never been seen in the ovary, testis, anterior mediastinum or cranial cavity.” We report here a case of intracranial fetus-in-fetu, with survival of the larger twin.

**Case Report**

This 6-week-old baby girl presented with enlarging head size, widened cranial sutures, and tense fontanels. There had been no complications during the pregnancy or delivery, and the infant had been feeding well since birth.

**Examination.** The baby was alert and capable of crying and moving all limbs with normal tone. There were no focal neurological deficits. The head circumference measured 42.5 cm (outside the 90th percentile on the head size/age curve). The sutures were wide and the fontanels full.

Skull x-ray films showed an irregularly rounded area of calcification centrally placed in the skull, with areas of translucency (Fig. 1). Computerized tomography (CT) scan (Fig. 2) showed severe supratentorial hydrocephalus, with the ventricular system dilated down to the third ventricle. There was no significant midline displacement. Within the posterior fossa, neural tissue was seen to extend as far as the foramen magnum, but it was difficult to delineate the cavity of the fourth ventricle. The appearance suggested caudal displacement of the cerebellum.

In the superior part of the posterior fossa, a large triangular cerebrospinal fluid (CSF)-filled space could be followed in continuity through the tentorial hiatus. The latter appeared abnormal in shape and was dilated. In the midline, supratentorially, was a large heterogeneous mass containing dense areas of calcification and fat. From this mass, isodense portions were seen to pass from the third ventricle into the anterior horns of the lateral ventricles, predominantly on the left side. Within these isodense portions, cystic areas could be seen.

**Operation.** A ventriculoperitoneal shunting procedure was performed first. A week later the patient was operated on in the face-down position, under general anesthesia, endotracheal intubation, and positive-pressure ventilation. A right-sided occipital scalp and bone flap was cut, extending the medial margin to the midline. Under the operating microscope, the occipital lobe was retracted from the falx without damaging any veins. At the posterior opening of the falx, a splayed-out cingulate gyrus could be seen instead of the splenium of the corpus callosum. A small incision was made into the right cingulate gyrus, the lateral
ventricle was entered, and the tumor was encountered. This appeared as a discrete mass with small hairs on its surface.

The tumor was very tough and a knife made little impression on it. Gentle traction of the exposed mass with pituitary rongeurs enabled a portion of it to be pulled through the cortical incision. At this stage it was evident that the surgeon was retracting on a limb with well-formed toes and rudimentary nails. With further gentle retraction and minimal dissection, an entire lower limb was delivered, followed by the opposite lower limb and the remainder of the mass.

Careful examination of the intraventricular area failed to show any definite attachment or blood supply, although a few capillaries were coagulated with a bipolar cautery near the anterior aspect of the third ventricular floor.

Pathological Examination. The tumor was a four-limbed mass, with digits at the end of each limb and nervous tissue indicating the cephalad end (Fig. 3 left). A body stalk arose from the abdomen and contained gut loops, some cystic cavities, and a cutaneous surface resembling genital tubercles. Figure 3 right shows the mass with the fourth limb in situ after removal of the body stalk and fixation. A meningeal covering of the "brain" can be clearly seen. A stained transverse section at the midtrunk level is shown in Fig. 4. As is usual in fetus-in-fetu, no structures resembling a heart, kidney, or gonad were seen.

Histopathologically, the brain showed evidence of cytoarchitectonic arrangement (Fig. 5 left). The spinal cord was flattened and appeared thicker ventrally at some levels. Organized areas of anterior horn cells were not seen, but ventrally placed dorsal root ganglia were found with nerves running distally from them (Fig. 5 right). Small bowel was found in the body stalk, and large bowel inside the body cavity. Lung tissue was well developed. Within one of the vesicular structures in the body stalk, an abrupt transition from mucus-secreting columnar epithelium to a stratified nonkeratinizing type was seen.

Using cell cultures established from tumor fibroblasts and lymphocyte cultures of the parents and host, it was established that the chromosome complement of the parents was 46XY and 46XX, respectively, and that of the tumor and host 46XX. Banding patterns did not help to establish the origin of the X chromosomes.

Postoperative Course. The child made a good recovery. She remained alert and bright and fed normally from the breast. She was discharged home 3 weeks after removal of the mass. Five months later, the head circumference was 42 cm (at the 50th percentile) and a repeat CT scan showed small ventricles (Fig. 6). Two months postoperatively the child was neurologically intact and appeared to have normal development for her age as judged by all routine pediatric and neurological tests.
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**FIG. 3.** *Left:* Posterior view of the mass. The nervous tissue tapers into a cord-like structure. Below the "buttocks" can be seen the body stalk. *Right:* Anterior view after fixation and removal of the body stalk. The fourth limb has been placed in its original site.

**FIG. 4.** Photomicrograph of the midtrunk section showing several gut loops. The spinal cord is replaced by a few strands of nervous tissue. H & E, × 0.5.
At 10 months the child continued to be extraordinarily well. The skull was completely normal and the anterior fontanel was represented by a depression that just admitted part of a fingertip. The child sat up at 8 months and could wave good-bye and clap her hands on request. She tended to roll and made swimming movements, but could not crawl at that time. She was visually alert and appeared to hear quite normally and was achieving all her milestones in the normal manner.

At 12½ months the child remained neurologically normal and was considered to be advanced for her age. She could pull herself up to the standing position. She is now 18 months old and has normal speech for her age. She walks unaided with a normal gait and all neurological and psychological tests show her to be a bright and intelligent child with no neurological deficits.

Discussion

Using Potter's classification of twin pregnancy (Table 1), a fetus-in-fetu is an example of heteropagus (B iii). If a descriptive definition that includes the need for evidence of axial development is adopted, as Willis' suggests, the lesion must have its origin at around the 15th day after fertilization when the presomite embryo consists of a bilaminar disc and will show the beginning of axial development. At this time, the primitive streak appears and gives rise to the intra-embryonic mesoderm. At its cephalic extremity Hensen's node forms, and at its center the inward movement of cells gives rise to the blastopore; this in turn forms the notochordal process which rapidly elongates
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TABLE 1
Classification of twin pregnancy

A. Dizygous gestation:
   i free dizygous twins
   ii conjoined dizygous tissues—tissue mosaicism

B. Monozygous gestation:
   i free monozygous twins
   ii joined monozygous twins (equal and symmetrical)—
      diplopagus
   iii unequal and asymmetrically joined twins—heteropagus

by caudal migration of Hensen's node. Axial development of fetus-in-fetu requires that an anomaly exists at this stage. Dorsal displacement of the developing "second axis" into the embryonic ectoderm, and hence the neural plate, may explain the finding of the fetus inside the lateral ventricle.

Adequate anatomical descriptions are rare in previously published reports. In 1975, Knox and Webb reviewed 18 of the 19 cases reported to that date; however, they did not comment on the case of Grant and Pearn or Breslau and Rindfleisch, although the description of the latter does not include a specific account of axial development. The report of Lord suggests that development had not proceeded beyond the 8th week, a finding endorsed by Grant and Pearn, but in our case this mark had probably been exceeded as the gut loops had partly returned to the "abdomen," and lung development was further advanced than would be expected in the first trimester.

In the case described by Kimmel, et al., axial development was shown in only two of the "five fetuses" included in a cerebral tumor in a 19-day-old hydrocephalic girl. The tumor was supratentorial and there were "extremities enough for five fetuses." Despite Willis' criticism of this report (at p 443), it is possible that this mass represented a fetus-in-fetu, although some anatomical details are tantalizingly absent.

Fetus-in-fetu is undoubtedly most common retroperitoneally and may present as an incidentally discovered abdominal mass or abdominal distension with raised intra-abdominal pressure, or cause pressure on surrounding viscera. No case has exhibited the presence of renal or gonadal tissue, or of a heart. The "aorta" in this case resembles the type seen in acardiac monsters and is typical of an unperfused large fetal artery.

References


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