

Intraventricular twin fetuses in fetu

Case report

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The authors report a rare case of multiple intracranial fetuses in fetu, fulfilling Willis' traditional criteria, which include an axial and appendicular skeleton with surrounding organized tissue. This case was ascertained from studies of a full-term female neonate who presented with ventriculomegaly. A CT scan showed intracranial calcifications that were suggestive of an axial skeleton. Her birth weight was 3.176 kg (50th–75th percentile), length was 52 cm (90th percentile), head circumference was 35 cm (50th–75th percentile), and Apgar scores were 7 and 8 at 1 and 5 minutes, respectively. Prenatal ultrasonography studies performed at 12 weeks and 5 days, and 19 weeks and 6 days revealed normal findings. A 37-week prenatal ultrasonography study showed ventriculomegaly and obstructive hydrocephalus, with a possible intracranial teratoma. Cranial imaging at birth with ultrasonography, CT and MR imaging, and MR angiography demonstrated 2 complex intraventricular masses with cystic, solid, and bony elements. A craniotomy with resection of the masses was performed at 3 months of age. The infant survived and is now 12 months old with some developmental progress. Two axial skeletons, with accompanying rib cage and extremities, including well-formed feet and toes, were noted. Both anencephalic structures had skin with hair, fat, skeletal and smooth muscle, and bony structures with bone marrow and focal areas of calcification. Multiple viscera were present and included thymus, bowel, stomach, salivary gland, kidney, adrenal gland, lung, and presumed adnexal structures. A diagnosis of fetuses in fetu was rendered. Chromosomal studies of the child and tissue from the 2 fetuses in fetu showed normal female karyotypes. A single nucleotide polymorphism array analysis from the proband infant and tissue from the 2 identified fetuses in fetu appeared to be genetically identical. These results are consistent with a monozygotic twin embryonic origin of the fetus in fetu tissue, which is a mechanism that has been suggested in previous reports in which karyotypes, blood types, and limited genetic loci have been studied. This is the first report of a rare example of intracranial intraventricular twin fetuses in fetu for which a genome-wide single nucleotide polymorphism assay has confirmed their genetic identity.

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KEY WORDS • fetus in fetu • intraventricular mass • twin • single nucleotide polymorphism microarray

FETUS in fetu is a rare phenomenon in which a discrete vertebrate fetiform mass is found within the body of a twin. The term “fetus in fetu,” also known as “cryptodidymus,” was first coined by Meckel in the 1800s.^{4,8,12,27} In 1896, Gould and Pyle¹⁰ described the condition as one in which “a man may be pregnant with his brother or sister” and “an infant may carry its twin.” The first official case report of fetus in fetu was described by Young³⁰ in 1809, in which he recounted an infant with a retroperitoneal mass that had human features and identifiable vertebrae.

Abbreviations used in this paper: ICP = intracranial pressure; SNP = single nucleotide polymorphism.

In the majority of cases, fetus in fetu occurs as an abdominal mass.¹⁴ To date, there have been only 15 reported cases of an intracranial fetus in fetu, 4 of which are published in the non-English language literature. Here, we report a unique case of twin intraventricular fetuses in fetu. Additionally, we used SNP microarray technology to confirm that the host and twin fetuses in fetu had identical genomic complements (monozygotic triplets).

This article contains some figures that are displayed in color online but in black and white in the print edition.

Case Report

History and Examination. This female girl of Vietnamese ancestry was delivered vaginally at 38 weeks' gestation to a 34-year-old gravida 1 mother and a 54-year-old father. The pregnancy was the result of a natural conception without any form of fertility enhancement. The maternal gestational weight gain was 30 lbs with borderline high blood glucose measurements. The family history was negative for consanguinity, hydrocephalus, congenital brain malformations, infant deaths, seizure disorders, multiple births, or other familial disorders. An ultrasonographic study performed at 37 weeks' gestation showed dilated lateral ventricles and possible aqueductal stenosis, with concerns for hydrocephalus versus a holoprosencephalic cyst. The baby had a birth weight of 3.176 kg (50th–75th percentile), length of 52 cm (90th percentile), head circumference of 35 cm (50th–75th percentile), and Apgar scores of 7 and 8 at 1 and 5 minutes, respectively. She had a spontaneous cry, good suck response, initially decreased truncal tone, a positive Moro reflex, and normal symmetrical flexion and extension of all extremities. Ophthalmological examination findings were normal. Her 2 × 3-cm anterior fontanel was full with no evidence of sutural splaying; the posterior fontanel was closed. A small cephalohematoma and 2 small areas of cutis aplasia over the left parietal scalp were noted. On her 2nd day of life, the baby had a high-pitched cry, intact extraocular movements, and normal tone.

Based on prenatal ultrasonography concerns, a head CT scan was obtained, demonstrating marked ventriculomegaly and a midline intraventricular soft-tissue mass with organized ossified structures suggesting a fetiform mass. A posterolateral soft-tissue swelling external to the calvaria was identified as a cephalohematoma. A subsequent MR imaging study delineated the anatomy of the midline fetiform mass and demonstrated rudimentary pelvic structures sandwiched by the anterior horns of the lateral ventricles, with more complex bony structures and soft tissues posteriorly.

Despite these radiographic findings, the baby remained neurologically normal with good suck response, conjugate eye movement, and no signs of elevated ICP during workup in the neonatal intensive care unit. The child was discharged home with her family with plans for resection when she was older and/or showed signs/symptoms of neurological decline. During outpatient follow-up, she exhibited small increases in her head circumference without signs of elevated ICP. However, at 3 months of age the child's head circumference measured well above the 97th percentile at 45 cm. The fontanel was tense, her eyes were slightly deviated downward, and she was more irritable and less consolable. It was also the first clinic appointment at which she had not gained weight. Follow-up imaging studies showed increased hydrocephalus and enlargement of the fetiform mass with interval development of soft tissues demonstrating fat attenuation surrounding the more developed pelvis (Fig. 1). The vascular supply was not well delineated, but the enlargement and development of fat suggested that the fetuses in fetu continued to mature over time.

Operation. A left frontal craniotomy was used to approach the left lateral ventricle. The ventricle was entered through the left middle frontal gyrus; a moderate amount of CSF egressed when the ventricle was entered. The first structure that was visible was a well-formed circular skin-covered mass with mature dark hair extending from its surface. Dissection around this mass demonstrated skin-covered shoulders and upper extremities. The hair-bearing mass, shoulders, arms, and fingers could not be coagulated with bipolar electrocautery and required monopolar electrocautery and disarticulation for removal. The mass caudal to this was not covered by skin, but it was hemorrhagic and had 2 distal deformed skin-covered extremities attached to it in anatomically correct locations consistent with lower extremities. This axial structure was in correct anatomical location for a thorax. Dissection continued around the thorax, with extremities being present at the most caudal end. These extremities were removed en bloc and resembled fetal legs with anomalies of the digits. The midline, thoracic, and hemorrhagic mass was removed in toto. It appeared to be a pelvis, bony spinal column, and rib cage. Attention was turned to the second fetal structure. A hairless head structure with an orifice was removed. Similar to the first fetus, the upper extremities were removed from the midline axial mass by using monopolar cautery. The lower extremities were coagulated and disarticulated from the thorax. As the second thorax was removed, large blood vessels to the fetus were identified from the posterior third ventricle and the top of the midbrain. These were cauterized using bipolar cautery and cut. The lower extremities were removed. Examination of the surgical cavity did not demonstrate residual masses. The frontal, temporal, and parietal cerebral cortices had completely collapsed onto the large surgical cavity; despite this, subdural bleeding was minimal. The space occupied by the mass, ventricle, and subdural spaces was filled with saline, and subdural and ventricular drains were left in place.

Postoperative Course. Immediately postoperatively, the patient remained intubated but had spontaneous respirations and vigorous movements of all limbs. On the 2nd postoperative day, the patient experienced a seizure and progressed to status epilepticus. A head CT scan revealed multiple foci of new bilateral intraparenchymal hemorrhages and no residual mass. An MR imaging study revealed similar findings, including continued subdural fluid collections, despite the presence of subdural drains (Fig. 2). The patient was placed in a pentobarbital coma for treatment of status epilepticus, which resulted in only incomplete control of her seizures. She also developed diabetes insipidus, which was treated aggressively. All the intracranial drains were eventually removed without elevated ICP.

After 3 weeks of intensive care, the family decided to change the patient's status to "do not resuscitate." She was extubated and remained on full medical and feeding management with comfort and supportive care service through palliative hospice care. She subsequently began to improve and was discharged home, tolerating full feeds and on a regimen of antiepileptic medication. At 6 months of age, her fontanel was full and her head circumference

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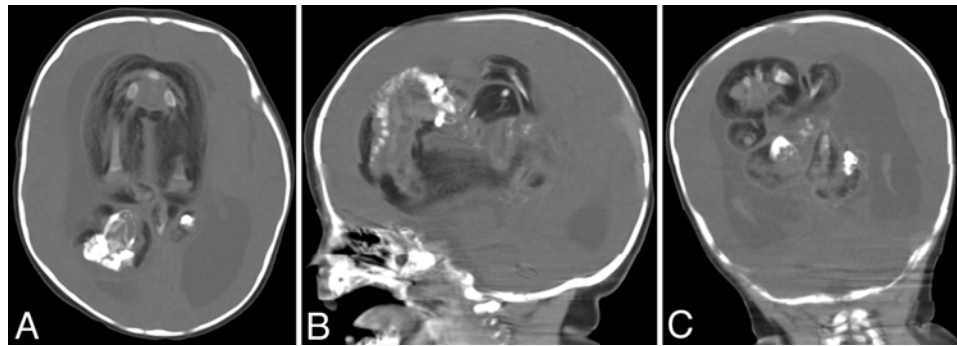


FIG. 1. Axial (A), sagittal (B), and coronal (C) head CT images showing 2 large intraventricular fetiform masses (fetuses in fetu) with obvious vertebral columns.

had grown to 47 cm, which was a significant increase compared with her immediate postoperative measurement of 44 cm. She was tracking with her eyes and had spontaneous symmetrical movement of all extremities, normal tone throughout, and a social smile. The patient could turn her head from side to side, but she had significant weakness in her neck muscles with a head lag. She did not use her hands, sit up, or make sounds. A head CT scan demonstrated the presence of cortical mantle in the parietal and occipital lobes only. The collapse of the frontal cortex was believed to be due to the chronic compression from the mass on an immature brain, as well as the mechanical trauma involved in the removal of the patient's intracranial mass and loss of CSF. A shunt was placed due to hydrocephalus, which led to a decrease in the head circumference. Follow-up head CT scans demonstrated improvement in the cortical mantle width. At the patient's 1-year postoperative follow-up, the child's head circumference showed stable growth along a slightly enlarged curve. She would smile and track to voice. She made eye contact and could babble. The child was taking most of her nutrition by mouth. However, her tone was poor throughout, with minimal head control; she did not sit and the beginning of spasticity was evident in her lower extremities.

Pathological Examination. Macroscopic evaluation of the surgical specimen revealed a 230-g aggregate (17.0 × 13.0 × 2.4 cm) of fetiform parts that included 2 anencephalic structures, one of which had an attached lower extremity. There were 3 additional lower extremities and 4 upper extremities (Fig. 3). All of the tissue fragments were covered with white-tan skin with lanugo-type hair. Both anencephalic structures had a rim of dark black hair. The cut surfaces were notable for 2 grossly identifiable vertebral columns. The extremities had from 4 to 7 recognizable digits and were composed of cartilaginous long bones with apparent joints and surrounding soft tissue.

Histologically, the fetiform parts were remarkable for skin with hair and adnexa, fat, skeletal and smooth muscle, and bony structures with bone marrow and focal areas of calcification. Two vertebral columns, an appendicular skeleton, and a bony jaw were noted, the latter containing identifiable tooth structures (Fig. 4A and B). Multiple viscera were present including thymus, bowel, stomach, salivary gland, kidney, adrenal, lung, and presumed adnexal structures (Fig. 4C and D). No definite brain or spinal cord tissue was identified, although nerve roots were seen at vari-

ous spinal cord levels. Likewise, no heart was identified by either gross or microscopic inspection.

Genetics Examination. Three days after birth, interphase fluorescence in situ hybridization was performed on uncultured peripheral lymphocytes obtained from the patient, targeting unique sequences on chromosomes 13 (RB1) and 21 (D21S342). These studies, as well as the conventional cytogenetic analysis, had results that were within normal limits (46,XX). Further cytogenetic analysis of tissue obtained from each of the fetuses in fetu also showed a normal female complement (46,XX).

The DNA from the patient and the fetuses in fetu was evaluated using an SNP microarray (6.0 Affymetrix Ge-

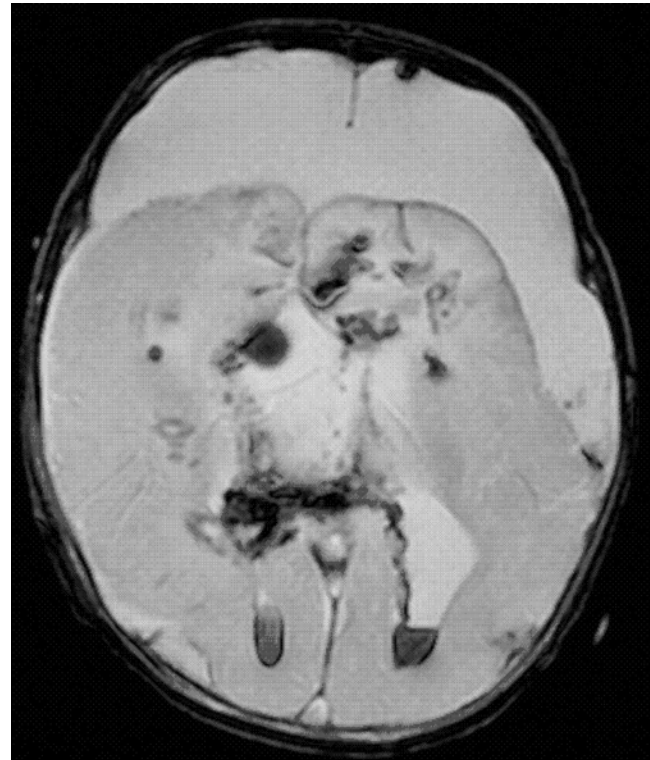


FIG. 2. Postoperative axial gradient echo MR image demonstrating thinned cerebral parenchyma that collapsed into the surgical cavity due to removal of the large fetiform mass. Areas of hypointense signal within the ventricles and scattered throughout the brain are indicative of multifocal intracranial hemorrhage.

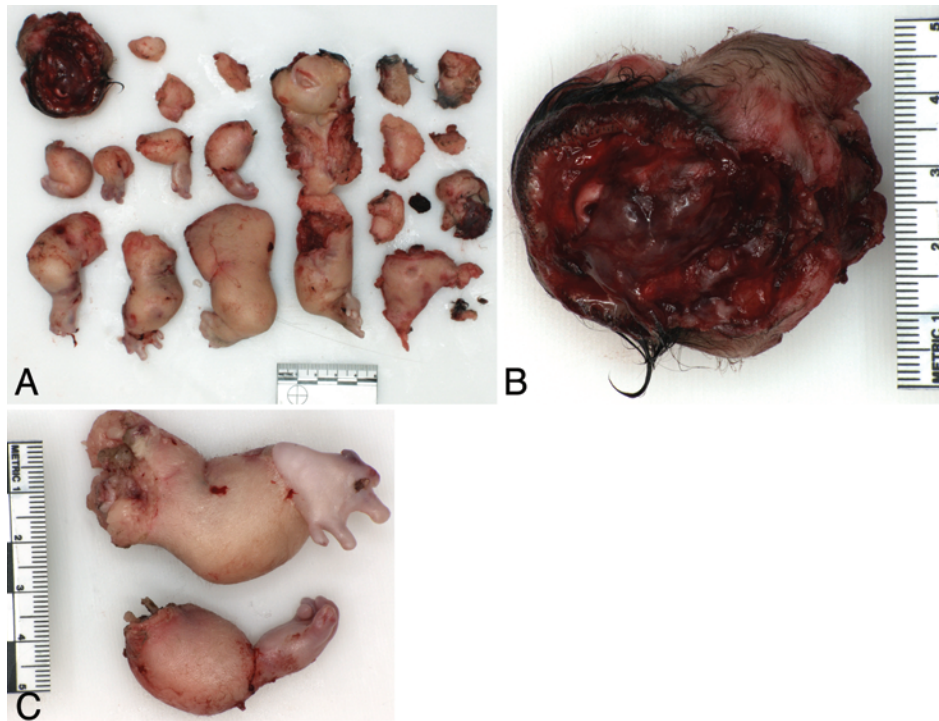


FIG. 3. Photographs. The gross appearance of the resected fetiform surgical specimen is shown in its entirety (A). An anencephalic head with lanugo-type hair covering the cutaneous surface (B) and upper and lower extremities with apparent long bones and from 4 to 7 digits (C) were among the tissue retrieved.

nome-Wide Human SNP). No significant DNA copy number changes were detected (using a 200-kb deletion/500-kb duplication threshold). Also, no increase in homozygosity (as one might see with uniparental disomy or consanguin-

ity) was noted. Importantly, a comparison of the SNP patterns in the host infant and fetal tissues showed identical genotypes, which is a finding consistent with monozygosity.

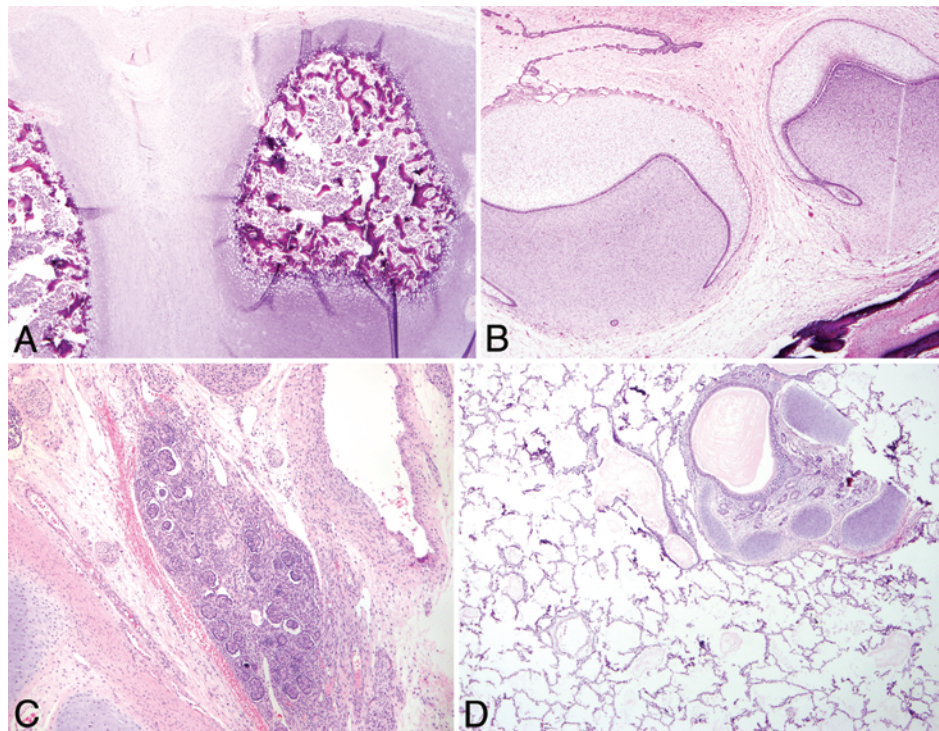


FIG. 4. Representative photomicrographs from the current case of twin fetuses in fetus, including vertebral structures (A), tooth buds (B), an immature kidney (C) and lung (D) tissue. H & E, original magnification $\times 200$ (A, B, and D) and $\times 400$ (C).

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Discussion

While the term “fetus in fetu” was coined and the first case was reported in the early 1800s, it was not until 1935 that Willis²⁸ proposed specific diagnostic criteria for this diagnosis. He stated that the presence of a vertebral column distinguished incomplete twins from a teratoma. In 1956, Lord¹⁹ expounded on this statement by requiring a vertebral axis, as well as “often ... an appropriate arrangement of other organs or limbs with respect to the axis” for a diagnosis of fetus in fetu. In contradistinction, Spencer²⁵ argued that the requirement of a vertebral column is too strict and that the mass need only be “enclosed within a distinct sac, partially or completely covered by normal skin, with grossly recognizable anatomic parts and attached to the autosite by a pedicle containing a few relatively large blood vessels.” While some accept Spencer’s less rigid definition of fetus in fetu, most authors adhere to the criteria set forth by Willis and Lord. The case reported here fully met the Willis and Lord criteria.

The incidence of fetus in fetu is estimated to be 1 in 500,000 births.¹¹ Due to the requirement of an axial skeleton, the etiology of this phenomenon is speculated to be an incomplete division of the blastocyst inner cell mass after formation of the primitive streak in monozygotic diamniotic monochorionic pregnancies.^{4,11,27} Given that monozygotic twins, conjoined twins, and our (and other) case(s) of twin fetuses in fetu are genetically identical, they may represent a continuum of completeness in the separation of one identical twin/triplet from the other. Prenatal diagnosis of fetus in fetu is rare, with ultrasonography studies showing only indistinct cystic and solid masses. The diagnosis is usually made through CT or MR

imaging studies obtained in the newborn, which was the experience with this patient.^{16,29}

The fetiform masses themselves are usually encapsulated in an amnion-like membrane and are often connected to the host by a vascular structure.^{4,12} To date, all reported cases of these masses have been anencephalic, and often the lower limbs have been better developed than the upper limbs. Histologically, tissues of the central nervous, gastrointestinal, and genitourinary systems are most prevalent. All cases of fetus in fetu studied thus far have been reported as acardiac.¹⁴ In 80% of cases, the mass has been located in the retroperitoneum, with the chief complaint usually being an abdominal mass.¹⁴ Fetus in fetu has also been reported in the mediastinum, sacrococcygeal area, kidneys, and oral cavity. Uncommon locations include the CNS, genitourinary system including scrotal placement, lungs, adrenal glands, pancreas, spleen, and lymph nodes.^{7,13}

There have only been 15 previously reported cases of intracranial fetiform masses^{1,5,6,9,13,15–18,20–24,26,29} (Table 1). The anatomical locations for these masses include posterior cerebrum/cranial fossa, supratentorial, temporooccipital sites, and “intracranial” not otherwise specified; 4 previous cases were reportedly intraventricular. In addition to these 15 cases, Gould and Pyle¹⁰ cited Aretaeos of Athens and his description of 3 fetuses within the cranial cavity of a 7-month gestational age twin.

When evaluating the number of fetiform structures in these intracranial cases, the reported number ranges from 1 to 7 structures (Table 1). It could be argued, however, that not all of these cases represent true a fetus in fetu, depending on which definition is applied. The shared patient reported on by Saito et al.²⁴ and Miura et al.²¹ was

TABLE 1: Published cases of intracranial fetus in fetu

Authors & Year	Language of the Report	Site	No. of Fetiform Structures	Vertebral Column Present?
Frank, 1903	German	unknown	unknown	unknown
Kimmel et al., 1950	English	posterior cerebrum	5	yes, in 2 of 5
Fiedler & Röse, 1974	German	unknown	unknown	unknown
Afshar et al., 1982	English	rt lat ventricle	1	“spinal cord”
Povýsilová, 1983	Czech	unknown	unknown	unknown
Yang & Leow, 1992	English	lt lat ventricle	1	yes
Tsai et al., 1993	Chinese	unknown	unknown	unknown
Hung & Lam, 1993	English	lt lat & 3rd ventricles	1	yes
Naudin ten Cate et al., 1995	English	posterior cerebrum	6	yes, in 6 of 6
Goldstein et al., 1996	English	“intracranial”	1	no
Ianniruberto et al., 2001	English	posterior cranial fossa	1	yes
Miura et al., 2006*	English	“intracranial”	1 fetiform mass & 6 additional masses	yes in fetiform mass; unknown in additional masses
Saito et al., 2007*	English	posterior cranial fossa	6	yes, no specified number
Kim et al., 2008	English	supratentorial	1	“notochord”
Heuer et al., 2008	English	temporooccipital	1	no
Marnet et al., 2008	English	3rd ventricle	2	no
present case	English	lt lat ventricle	2	yes, in 2 of 2

* These case reports are of the same patient.

described as having 6 or 7 masses, but apparently only the largest structure contained limbs and a vertebral column, and thus was the only mass that was definitively a fetus in fetu. A vertebral column was similarly not identified in several cases; thus, they did not conform to the criteria set forth by the definition of Willis²⁸ or Lord¹⁹ of fetus in fetu. Goldstein et al.⁹ reasoned that the well-developed limbs in their case argued for the diagnosis of fetus in fetu, even without the presence of a vertebral column. The case reported by Heuer et al.¹³ was that of an “ulnalike bone and pseudoarticulations with the right calvarium.” Marnet et al.²⁰ described a “non-vertebrate mass covered by normal skin” and argued that the histology of organized squamous epithelium with skin appendages qualifies under the description of fetus in fetu by Spencer.²⁵

The current case of intraventricular twin fetuses in fetu meets Willis’ traditional criteria²⁸ and has many of the same findings as previously described cases. Similar to previous cases, the diagnosis was not made until after birth. A vascular pedicle connected the 2 masses to the host’s circulation. Both fetiform structures were anencephalic with well-formed limbs, which were better developed in the lower extremities. A vertebral column was present in each structure. In addition to CNS, gastrointestinal, and genitourinary tissues, less common organs such as thymus, salivary gland, tooth buds, and adnexal structures were noted. While several previously reported cases of CNS fetus in fetu have described more than 1 fetal mass, many of these cases did not have a vertebral column. Kimmel et al.¹⁸ did describe 2 vertebrate structures, but it is not definitive that they were found in the ventricle. Adhering to the criteria set forth by Willis and Lord, our case therefore appears to represent the first reported case of intraventricular twin fetuses in fetu.

The SNP array analysis performed on tissue from this infant and tissue from the fetuses in fetu structures confirmed that all were genetically identical, which is consistent with a multiclave embryo. This supports the monozygotic theory of mechanism of origin for fetuses in fetu and may represent aberrant monozygotic triplet formation in our current case. This finding, which is the first to use SNP microarray technology to determine zygosity status, is in agreement with the results of previous investigators who determined zygosity by using chromosomal heteromorphisms, blood type polymorphisms, multiple tandem repeat polymorphisms (limited to 5–10 marker loci), or protein polymorphisms.^{2,3,17,21} All reports that included genetic analyses have shown that the fetus in fetu and the host were genetically identical.

In summary, the current case represents a unique example of intracranial, and more specifically intraventricular, monozygotic twin fetuses in fetu. It represents the only reported case of a multiple gestation within the intraventricular system of the CNS that meets the traditional criteria for fetus in fetu, with an associated axial skeleton, chest cavity, and surrounding organ formation.

Disclosure

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Author contributions to the study and manuscript preparation include the following. Conception and design: Huddle, Fuller, Tye, Ritter. Acquisition of data: Huddle, Fuller, Powell, Hiemenga, Yan, Deuell, Lyders, Papenhausen, Tye. Analysis and interpretation of data: Huddle, Fuller, Powell, Hiemenga, Yan, Deuell, Lyders, Papenhausen, Jackson-Cook. Drafting the article: Huddle, Fuller, Powell, Hiemenga, Yan, Deuell, Lyders. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Huddle. Administrative/technical/material support: Papenhausen.

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