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How Accurately Does Current Fetal Imaging Identify Posterior Fossa Anomalies?

Catherine Limperopoulos^{1,2,3}, Richard L. Robertson Jr.⁴, Omar S. Khwaja¹, Caroline D. Robson⁴, Judy A. Estroff⁴, Carole Barnewolt⁴, Deborah Levine⁵, Donna Morash⁶, Luanne Nemes⁶, Linda Zaccagnini⁶, and Adré J. du Plessis¹

¹*Fetal-Neonatal Neurology Research Program, Department of Neurology, Children's Hospital Boston and Harvard Medical School, Boston, MA.*

⁴*Department of Radiology, Children's Hospital Boston and Harvard Medical School, Boston, MA.*

⁵*Department of Radiology, Beth Israel Deaconess Medical Center and Harvard Medical School, Boston, MA.*

⁶*Advanced Fetal Care Center, Children's Hospital Boston and Harvard Medical School, Boston, MA.*

Abstract

OBJECTIVE—The first objective of our study was to describe the prevalence and spectrum of posterior fossa anomalies over 5 years in a major fetal care center where the referral diagnosis (by fetal sonography) was investigated by fetal MRI and, if confirmed, by postnatal MRI if possible. The second objective was to assess the accuracy with which fetal MRI predicts postnatal MRI findings in this population.

MATERIALS AND METHODS—We retrospectively identified all cases of suspected fetal posterior fossa anomalies referred to our center from 2002 through 2006. We reviewed maternal, fetal, neonatal, and follow-up records of all cases and fetal and early postnatal imaging studies.

RESULTS—Of the 90 cases of suspected fetal posterior fossa anomalies (by fetal sonography) referred over the study period, 60 (67%) were confirmed by fetal MRI. Of 42 live-born infants, 39 (93%) underwent postnatal MRI. There was complete agreement in fetal and postnatal MRI diagnoses in 23 infants (59%). In 16 cases (41%), fetal and postnatal MRI diagnoses disagreed; postnatal MRI excluded fetal MRI diagnoses in six cases (15%) and revealed additional anomalies in 10 cases (26%).

CONCLUSION—Although a valuable adjunct to fetal sonography in cases of suspected posterior fossa anomaly, current fetal MRI, particularly in early gestation, has limitations in accurately predicting postnatal MRI abnormalities. Advancing the accuracy of MRI for the diagnosis of posterior fossa anomalies will require greater understanding of normal brain development and improved tissue resolution of fetal MRI. During the interim, our findings strongly support the need for postnatal MRI follow-up in cases with suspected posterior fossa anomalies by fetal MRI.

Keywords

brain development; cerebellum; fetal MRI; fetal sonography; obstetrics; pediatric imaging; posterior fossa anomalies

Posterior fossa malformations are among the most common brain anomalies identified by current fetal imaging techniques. Advances in MRI during fetal and early postnatal life continue to provide important insights into normal and abnormal development of the cerebellum and brainstem [1–6]. Despite the rapid progress in fetal imaging, the prenatal diagnosis of posterior fossa dysgenesis remains challenging due to both false-positive and false-negative diagnoses [7–13]. As a result, a variety of classification schemes have been proposed for posterior fossa anomalies [14–17] without any scheme receiving widespread acceptance to date.

Abnormalities of posterior fossa development can, in general, be divided into those in a small posterior fossa with “crowding” of its contents (e.g., Chiari’s malformations) versus those occurring in a normal or enlarged posterior fossa. The latter category, which is the focus of this study, may be further subcategorized into anomalies in a posterior fossa with normal fluid spaces versus those associated with enlarged (relatively or absolutely) posterior fossa fluid spaces.

The primary reason for performing fetal imaging is to gather accurate information about fetal structure on which to base reliable counseling. This counseling to parents is important both for obtaining appropriate knowledge of the potential outcomes in the neonate and child and for making informed decisions regarding future management of the pregnancy. Although a number of studies have compared the findings of fetal sonography and postnatal MRI [11,13,18,19] and of fetal imaging versus pathology results [8,10,20,21], there is a paucity of data comparing the findings on MRI performed during the fetal period versus postnatal MRI in pregnancies that result in delivery of live-born infants.

Several objectives were the focus of this study. First, we aimed to describe the prevalence and spectrum of posterior fossa anomalies on sonography and MRI in a major referral center for fetal care over a 5-year period. Second, we wanted to determine the value of performing fetal MRI studies when posterior fossa anomalies are diagnosed on fetal sonography. Third, we chose to assess the accuracy with which fetal MRI predicts postnatal MRI findings in this population. Finally, we began to explore potential reasons for disagreement between fetal and postnatal studies. This study is intended to be viewed from the vantage point of clinicians who depend on the reliability of prenatal diagnosis and accuracy of interpretation during the difficult counseling of parents with a fetus in whom a posterior fossa anomaly is suspected.

Materials and Methods

Selection Criteria and Procedures

We retrospectively identified all pregnant women referred for counseling for suspected fetal posterior fossa anomalies at our center from 2002 through 2006. We excluded cases of suspected posterior fossa crowding, such as Chiari’s malformations with or without spinal dysraphism.

Our center is a regional referral center evaluating approximately 500 pregnancies per year for fetal abnormalities, of which approximately 20% are for suspected CNS anomalies. A dedicated multidisciplinary team of radiologists, neuroradiologists, neurologists, and neurosurgeons with expertise in fetal CNS anomalies performs all fetal consultations for suspected CNS anomalies. All cases of suspected fetal CNS anomaly undergo fetal MRI. All fetal imaging studies are reviewed by specialist fetal radiologists, with additional consultation from pediatric neuroradiologists in all cases of fetal CNS anomalies.

All consultations with expecting parents occur in the center. Patients are then referred back to their primary obstetrician or maternal–fetal specialist with the fetal diagnosis and a plan for follow-up. Specifically, at the initial consultation arrangements are made for the postnatal

follow-up of all surviving infants in our program staffed by the same neurologists who perform the fetal consultations. The program maintains a detailed database of all clinical visits. Children with suspected CNS anomalies in our program undergo postnatal MRI during early infancy and as indicated clinically thereafter. The committee on clinical investigation at our institution approved this retrospective study.

Medical Record Review

We reviewed maternal, fetal, neonatal, and follow-up records of all cases of suspected posterior fossa anomalies referred over a 5-year period. From this review, we recorded the dates of fetal and postnatal MRI examinations, imaging diagnoses (both inside and outside the CNS), gestational age at prenatal MRI (based on last menstrual period or, if unknown, on earliest fetal sonography), fetal sex, maternal age, parity, singleton versus multiple gestation, fetal karyotype (when available), gestational age at birth, birth weight, age at postnatal MRI, and additional postnatal genetic information. We also established whether pregnancy was terminated electively and documented the calendar year of prenatal MRI. Of note, in this study, we use the diagnostic terms used by the referring community obstetricians as the initial fetal sonographic diagnosis.

Prenatal and Early Postnatal Neuroimaging Techniques

Prenatal MRI studies were performed at Children's Hospital in Boston, MA, using a 1.5-T TwinSpeed Signa system (GE Healthcare) and an 8-channel phased-array cardiac coil. Multiplanar single-shot fast spin-echo (SSFSE) T2-weighted imaging sequences were performed using the following parameters: TE_{eff} , 80 milliseconds; number of excitations (NEX), 0.5; field of view, 40×40 cm; section thickness, 4 mm with no interslice gap; and acquisition matrix, 256×256 . At Beth Israel Deaconess Medical Center, prenatal imaging was also performed using a 1.5-T Twin-Speed Signa system (GE Healthcare) with a 4- or 8-element surface coil and a similar SSFSE imaging sequence with the following parameters: TE_{eff} , 90 milliseconds; NEX, 0.5; field of view, 26×30 cm; slice thickness, 4 mm with no interslice gap; and acquisition matrix, 256×256 .

Multiplanar SSFSE T1-weighted imaging sequences were performed at both institutions using the following parameters: TR/ TE_{eff} , 2,500/1.4; inversion time, 2,000 milliseconds; NEX, 1; field of view, 40×40 cm; section thickness, 4 mm with no interslice gap; and acquisition matrix, 256×192 .

All postnatal imaging studies were performed at Children's Hospital Boston using a quadrature or 8-channel phased-array head coil. Sagittal and axial spin-echo T1-weighted sequences (TR/TE, 600/20; NEX, 2; field of view, 20×20 cm; section thickness, 4 mm with 1-mm gap; and acquisition matrix, 256×192) and axial fast spin-echo T2-weighted sequences (3,200/85; NEX, 1; field of view, 20×15 cm; section thickness, 4 mm with 1-mm gap; acquisition matrix, 256×192 ; echotrain length, 8) were performed. Susceptibility-sensitive imaging was performed using a multiplanar gradient recall gradient-echo technique (600/40; NEX, 1; field of view, 20×20 cm; section thickness, 4 mm with 1-mm gap; flip angle, 30° ; acquisition matrix, 256×256).

Neuroimaging Diagnostic Criteria

Dandy-Walker malformation was diagnosed when the following three criteria were met: first, vermian agenesis or hypogenesis; second, cystic dilatation of the fourth ventricle; and, third, an abnormally high tentorium with enlargement of the posterior fossa. We, like others [22], avoid using the term "Dandy-Walker variant" because the inconsistent application of this term has severely limited its utility [22]. Inferior vermian hypoplasia was diagnosed when caudal growth of the inferior vermis over the fourth ventricle remained incomplete after 18–20 weeks

of gestation, as assessed on MRI in the midline sagittal plane. The term “isolated inferior vermian hypoplasia” was used to indicate that this lesion was present with normal-shaped or near-normal-shaped cerebellar hemispheres, a normal-sized posterior fossa without obvious cystic lesions, and normal supratentorial structures. Cerebellar hypoplasia was diagnosed when one or both cerebellar hemispheres were small with short but normally arranged fissures. A diagnosis of rhombence-phalosynapsis was made when the vermis was absent and midline fusion of the two cerebellar hemispheres was detected. A diagnosis of mega cisterna magna corresponded to an enlarged retrocerebellar space (> 10 mm), presumably due to a variant in skull growth, with an otherwise normal cerebellum, with normal bulging of the pons, with normally inserted tentorium, and without ventricular enlargement. A retrocerebellar arachnoid cyst was diagnosed when a cystic pouch behind the cerebellum was not in communication with the fourth ventricle. Cerebellar hemorrhage was defined as areas of increased signal seen in the cerebellum on T1-weighted images.

After all cases of posterior fossa anomaly were identified, we assessed the agreement between the referring sonography diagnosis and the fetal MRI diagnosis. We compared the fetal MRI diagnosis with postnatal MRI studies and examined the association between discrepancies in pre- and postnatal MRI and the gestational age at fetal MRI, calendar year of fetal MRI, and type of posterior fossa anomaly.

Statistical Analysis

Continuous perinatal data were summarized using the median, mean, and SD, and categorical factors were summarized using proportions. Group differences on clinical and neuroimaging variables were compared with the Student's *t* test or Wilcoxon's rank sum test for continuous variables or the Fisher's exact test for dichotomous data. For subgroups of infants categorized by posterior fossa diagnoses, differences in continuous variables were evaluated using one-way analysis of variance; ordinal variables were compared using the Kruskal-Wallis test and categorical variables using Fisher's exact test. Additional analyses controlling for gestational age at fetal MRI, calendar year of fetal MRI, and type of posterior fossa anomaly were performed using multiple linear and logistic regression analyses.

Results

Characteristics of the Study Cohort

Over the 5-year study period, 90 pregnant women were referred for suspected fetal posterior fossa anomalies (15 in 2002, 16 in 2003, 11 in 2004, 25 in 2005, and 23 in 2006). Mean maternal age ($\pm$ SD) was 30.5 ± 5.3 years (range, 17–41 years). Forty-six percent of the women were primiparous. Eighty-eight of the pregnancies (98%) were singleton; in each of two twin pregnancies, only one fetus was referred for suspected fetal posterior fossa anomalies. Of the 90 fetal cases, 55 were male, and 26 were female; sex was unspecified in nine fetuses. The outcomes for all posterior fossa anomalies identified over the 5-year study period are summarized in Figure 1.

Fetal MRI Findings

Of the 90 cases referred for suspected posterior fossa anomalies by fetal sonography, 25 (28%) were found to have normal posterior fossa structures by fetal MRI (Table 1) and five were found to have supratentorial ventriculomegaly only. In the remaining 60 cases (67%), posterior fossa abnormalities suspected by fetal sonography were confirmed by fetal MRI. These abnormalities were inferior vermian hypoplasia ($n = 22$ [37%]), of which 18 were isolated inferior vermian hypoplasia, Dandy-Walker malformation ($n = 15$ [25%]), mega cisterna magna ($n = 9$ [15%]), arachnoid cyst ($n = 5$ [8%]), cerebellar hemispheric (unilateral or bilateral) hypoplasia ($n = 4$ [7%]), cerebellar hemorrhage ($n = 4$ [7%]), and

rhombencephalosynapsis ($n = 1$ [2%]). Concomitant supratentorial anomalies were present in 19 cases (32%) in which fetal MRI showed posterior fossa abnormalities, as shown in Table 2.

Fourteen cases (23%) underwent follow-up fetal MRI study at a mean gestational age of 33.6 ± 2.5 weeks (range, 29–36 weeks) and an interval range between the first and second fetal MRI examinations of 7–10 weeks. Diagnostic categories of repeat fetal MRI consisted of Dandy-Walker malformation ($n = 5$), ventriculomegaly ($n = 4$), arachnoid cyst ($n = 2$), inferior vermian hypoplasia ($n = 2$), and cerebellar hemispheric hypoplasia ($n = 1$). Additional CNS findings documented on follow-up fetal MRI studies included agenesis of the corpus callosum in four cases with Dandy-Walker malformation and ventriculomegaly, and cerebral heterotopias in two cases, one with arachnoid cyst and one with inferior vermian hypoplasia.

Associated Extracerebral Anomalies

Fourteen of 60 cases (23%) with fetal posterior fossa abnormalities diagnosed by fetal MR also had associated extracerebral anomalies including cardiac malformations ($n = 3$), ambiguous genitalia ($n = 2$), renal dysplasia ($n = 2$), cardiac malformation and echogenic kidneys ($n = 1$), echogenic bowel ($n = 1$), vertebral anomalies ($n = 1$), extremity malformations ($n = 1$), hypertelorism and micrognathia ($n = 1$), echogenic bowel and hypertelorism ($n = 1$), and congenital diaphragmatic hernia ($n = 1$).

Pregnancy Outcomes

Of the 60 cases of posterior fossa abnormality by fetal MRI, 36 women underwent amniocentesis, which revealed chromosomal abnormalities in four fetuses (trisomy 18 in two, partial trisomy 13 in one, and Smith-Lemli-Opitz syndrome in one). Of the 60 pregnancies with posterior fossa abnormalities detected on fetal MRI, 18 pregnancies (30%) were terminated electively (Table 3). Of the remaining 42 pregnancies, all fetuses were born alive, at a mean gestational age of 37.6 ± 3.5 weeks (range, 26–42 weeks) and mean birth weight of $2,954.9 \pm 1,156.9$ g (range, 920–4,500 g). Five infants died in the early postnatal period with diagnoses of Dandy-Walker malformation, hydrocephalus, and agenesis of the corpus callosum ($n = 2$); Dandy-Walker malformation and trisomy 18 ($n = 1$); isolated inferior vermian hypoplasia, trisomy 18, and congenital diaphragmatic hernia ($n = 1$); and isolated inferior vermian hypoplasia with hypoplastic left heart syndrome ($n = 1$).

Agreement Between Fetal and Postnatal MRI Diagnoses

Of the 42 live-born infants, 39 (93%) underwent postnatal MRI at a mean age of 3.2 ± 4.9 months (range, 1 week–15 months). There was complete agreement between the fetal and postnatal MRI diagnoses in 23 infants (59%). Diagnostic categories among these 23 cases included isolated inferior vermian hypoplasia ($n = 8$), cerebellar hemorrhage ($n = 4$), Dandy-Walker malformation ($n = 4$), mega cisterna magna ($n = 3$), Dandy-Walker malformation and agenesis of the corpus callosum ($n = 2$), arachnoid cyst and cerebral heterotopias ($n = 1$), and arachnoid cyst ($n = 1$).

In the remaining 16 infants (41%), discrepancies in posterior fossa diagnoses were noted between the fetal and postnatal MRI studies and included cases of both over- and under-diagnosis in the fetal studies. In six cases (15%), posterior fossa anomalies diagnosed by fetal MRI could not be confirmed by postnatal MRI. Cases with discrepant findings between fetal and postnatal MRI studies are summarized in Table 4. Diagnostic categories of infants with normal findings on postnatal MRI included inferior vermian hypoplasia in five infants and mega cisterna magna in one infant. Conversely, in 10 cases (26%), postnatal MRI revealed additional supratentorial malformations and more extensive cerebellar anomalies not evident on the fetal studies (Fig. 2).

Predictors of Discrepant Fetal and Postnatal MRI Diagnoses of Posterior Fossa Anomalies

Lower mean gestational age (24.2 ± 4.5 vs 27.2 ± 4.8 weeks, $p < 0.01$) at the time of the fetal MRI was associated with a greater likelihood of discrepancy between fetal and postnatal MRI findings. Over the 5-year study period, there was a significant overall decrease in the proportion and type of fetal–postnatal MRI diagnostic discrepancies ($p = 0.02$). Examination of the type of posterior fossa lesions revealed a borderline trend for improved fetal MRI diagnostic accuracy for subtle lesions, such as isolated inferior vermian hypoplasia, mega cisterna magna, and cerebral heterotopias ($p = 0.051$).

Genetic Findings

Among infants with postnatal MRI confirmation of a posterior fossa anomaly, genetic syndromes and chromosomal abnormalities were identified by amniocentesis or postnatal testing in 25% (13/51) of the cases tested. These abnormalities included trisomy 18 ($n = 2$), partial trisomy 13 ($n = 1$), Smith-Lemli-Opitz syndrome ($n = 1$), *AH11* mutation ($n = 1$), PHACES syndrome (posterior fossa malformations; facial hemangiomas; arterial anomalies; cardiac anomalies and aortic coarctation; eye anomalies; and sternal clefting, supraumbilical raphe, or both) ($n = 1$), Beckwith-Wiedemann syndrome ($n = 1$), Rubinstein-Taybi syndrome ($n = 1$), Aicardi's syndrome ($n = 1$), cri du chat syndrome ($n = 1$), Wolf-Hirschhorn syndrome ($4p-$) ($n = 1$), partial trisomy 8p + monosomy 6q ($n = 1$), and 46 XXX ($n = 1$).

Discussion

Imaging techniques that allow diagnosis of fetal malformations have had a major impact on the management of pregnancy and of the neonate and infant, no more so than in cases of suspected fetal brain anomalies. Sonography has for some time been the primary fetal imaging technique and remains the screening technique of choice. However, fetal sonography has limited specificity, and false-positive diagnoses of posterior fossa anomalies are well described [9–11,13]. In our study, fetal MRI added diagnostic specificity for the diagnosis of posterior fossa anomalies when compared with screening fetal sonography in the community, with approximately 30% of such diagnoses by fetal sonography being excluded by fetal MRI. However, despite the apparent strengths of fetal MRI, neuropathology studies have raised concerns about the accuracy of fetal MRI for the diagnosis of brain anomalies [8,21]. The populations in these pathology studies are likely biased toward severe cases that undergo fetal autopsy. Few studies have compared the accuracy of fetal MRI with that of postnatal MRI for the diagnosis of brain malformations in general [23–25].

In our study, the first to our knowledge focusing on fetuses with posterior fossa anomalies surviving pregnancy, fetal MRI showed limitations in both sensitivity and specificity when compared with postnatal MRI. The accuracy with which fetal MRI predicted findings on postnatal MRI in cases of posterior fossa anomalies was modest at best, with only 60% of prenatal diagnoses confirmed postnatally.

There are several possible reasons for the limited ability described in this study of fetal MRI diagnosis of posterior fossa lesions to predict findings on postnatal MRI. The use of MRI for imaging the fetus is relatively new, at least in the United States, and although it represents a major advance in fetal diagnosis, many technical challenges remain, including limited pulse sequence options and low signal-to-noise ratios. In recent years, advances in MRI technology, such as faster scanning times, have improved but have not yet overcome the challenges of tissue resolution of in utero structures. Furthermore, although significantly reduced by ultrafast MR techniques, fetal motion artifact may still affect image quality and the ability to obtain a true sagittal image.

The issue of tissue resolution is particularly limiting for small structures such as the brainstem and vermis. This limitation of MRI was evident in our study, in which one quarter of the infants undergoing postnatal MRI had “new” posterior fossa lesions detected including brainstem hypoplasia, superior vermian dysplasia, and posterior fossa hemorrhage. In addition, small cerebral lesions (e.g., heterotopias) not noted on fetal MRI but seen on postnatal images may have major prognostic importance. These lesions may be too small to diagnose by MRI early in gestation. Conversely, in 15% of our cases, fetal MRI diagnoses could not be confirmed by postnatal MRI. Of note, in almost all cases in which a fetal posterior fossa anomaly was excluded by postnatal MRI, the fetal diagnosis was isolated inferior vermian hypoplasia. These findings corroborate our recent report that isolated inferior vermian hypoplasia may be overdiagnosed by second-trimester fetal MRI [7]. Similarly, a recent study comparing fetal MRI diagnosis of posterior fossa anomalies with results at fetal autopsy showed that vermian hypoplasia diagnosed by fetal MRI could not be confirmed by neuropathology studies in 30% of the cases [21]; of note, these false-positive MRI diagnoses were made after 30 weeks’ gestation [21]. In our study, one third of terminated pregnancies had isolated inferior vermian hypoplasia. Together, these findings suggest that caution is warranted during counseling for inferior vermian hypoplasia, particularly if the lesion is an isolated finding, because a good outcome is to be expected in the majority of cases [7].

Beyond the technical limitations of in utero tissue resolution by MRI, there are a number of important factors that likely underlie the discrepancies between fetal and postnatal MRI in cases of posterior fossa anomalies. One major limitation is our incomplete understanding of the normal temporal and structural range of fetal brain development, which may compromise reliable interpretation of the more subtle posterior fossa anomalies. Concerns regarding the limited resolution of fetal MRI in the posterior fossa have been raised by others, showing a delay of up to 5 weeks between known anatomic stages of development [26] and those detected by fetal MRI [27]. In our study, early gestational age at the time of fetal MRI was a significant predictor of discrepancy between the fetal and postnatal MRI posterior fossa findings. This is not surprising when considering the protracted time line of cerebellar development. Although the cerebellum is one of the first brain structures to begin development, at around 4 weeks of gestation, it is one of the last to reach its mature configuration, many months after birth [28]. In fact, during the middle and late fetal periods, the cerebellum undergoes particularly rapid changes in size and differentiation [1,2,8,28]. For example, cerebellar transverse diameter doubles in size during the second half of gestation [29]. The rapid growth is the result of important developmental processes that may be vulnerable to genetic and environmental insults capable of derailing the developmental program.

In our study, the likelihood of discrepancy between fetal and postnatal MRI studies decreased over the 5-year study period. Although the interpretation of fetal studies at our center is performed by experienced fetal ultrasonologists and by pediatric neuroradiologists in cases of suspected brain anomaly, the advent of fetal MRI at our center is relatively recent. Therefore, improved correlation between fetal and postnatal studies may possibly be in part due to the accumulation over time of experience in the application and interpretation of fetal MRI.

Given the critical role of brain function in long-term quality of life, the interpretation of fetal brain structure by MRI is often a pivotal determinant in difficult parental decisions about the future of the fetus when a fetal anomaly is suspected. Previous studies have described the current limitations of MRI in the diagnosis of fetal brain anomalies [7,30,31], and our study supports these findings in cases of suspected fetal posterior fossa abnormalities. Clinicians are often called on to make accurate predictions of fetal outcome against a background “ticking clock” for legal termination of pregnancy, despite the technical limitations of fetal MRI at this stage of gestation when many important developmental events are yet to occur. Our current and previous [2,7] findings should be carefully considered when predicting postnatal brain

structure by MRI in cases of suspected fetal posterior fossa abnormality. Furthermore, unlike the data of other investigators [23,32], our data strongly emphasize the importance of performing postnatal MRI studies in all cases of posterior fossa anomaly suspected by fetal MRI.

Our study has several potential limitations. First, the overall goal of the study was to examine—from the counseling physician's perspective—the current diagnostic accuracy in a major referral center of fetal MRI for posterior fossa anomalies. For this reason, we did not perform repeated blinded MRI interpretations to assess intra- and interreader reliability. Instead, we used the clinical diagnosis made by our pediatric neuroradiologists at the time of the prenatal and postnatal MRI studies. Second, given the large number of heterogeneous diagnostic groups in our cohort, our data did not have sufficient power to allow us to pursue more targeted analyses to evaluate the diagnostic accuracy of every posterior fossa lesion. In addition, because the cases with normal findings on fetal MRI studies did not undergo postnatal MRI studies, we are unable in this retrospective study to comment on the specificity of fetal MRI. Finally, this study focused on the structural diagnostic accuracy of fetal MRI and did not address the predicted and actual long-term functional outcomes. Those goals are part of a current ongoing study.

In conclusion, antenatal MRI has advanced our ability to define major posterior fossa anomalies in the fetus. However, from the perspective of the clinician counseling parents during the gestational period when critical decisions are often made, important limitations of the technique and its interpretation persist and need to be considered carefully. Our findings strongly support the need for postnatal MRI follow-up in cases with suspected posterior fossa anomalies by fetal MRI. Overcoming the current limitations of fetal MRI in this population will require not only further developments in the technique but also greater understanding of the temporal and structural variations in the development of posterior fossa structures. Such advances in our understanding will be facilitated by large prospective studies using serial and quantitative MRI in both the healthy fetus and in the fetus with suspected posterior fossa abnormalities.

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Fig. 1.

Summary of outcomes for all posterior fossa anomalies identified over 5-year study period.

Fig. 2.

Postnatal MRI confirms fetal MRI findings and shows additional findings not apparent on fetal MRI.

A and B, Single-shot fast spin-echo T2-weighted images of fetus at 21 weeks' gestation show hypoplasia of vermis and corpus callosum (**A**) and right cerebellar hemisphere (**B**).

C, Coronal T2-weighted image of 2-day-old female neonate confirms findings shown on **A** and **B**; however, in addition, postnatal MRI study shows thickened dysmorphic cerebellar cortex and right occipital subependymal nodular heterotopia not apparent on fetal MRI.

TABIE 1Fetal sonography Diagnoses of Posterior Fossa abnormalities Excluded by Fetal MRI ($n = 25$)

Referring Sonography Diagnosis (no. of cases)	Fetal MRI	
	Second Trimester (14–26 wk)	Third Trimester (27–39 wk)
Mega cisterna magna (7)	1	6
Dandy-Walker variant (7)	6	1
Inferior vermian hypoplasia (5)	5	0
Posterior fossa cyst (3)	2	1
Dilated fourth ventricle (1)	1	0
Enlarged cerebellum (1)	0	1
Ventriculomegaly, Dandy-Walker malformation (1)	1	0

Note—Data are number of cases.

TABLE 2

Posterior Fossa anomalies and associated supratentorial Findings on Fetal MRI ($n = 19$)

Posterior Fossa Anomaly	Supratentorial Findings	No. of Cases
Dandy-Walker malformation	ACC	3
	Ventriculomegaly	2
	ACC and ventriculomegaly	2
	ACC and brainstem hypoplasia	1
	ACC and cerebral heterotopias	1
Inferior vermian hypoplasia	Ventriculomegaly	2
	ACC	1
	Holoprosencephaly	1
Mega cisterna magna	Ventriculomegaly	1
	ACC	1
Cerebellar hypoplasia	ACC	1
	Cerebral heterotopias	1
Arachnoid cyst	Ventriculomegaly	1
	Cerebral heterotopias	1

Note—ACC = agenesis of the corpus callosum.

TABIE 3

Characteristics of Fetuses with Posterior Fossa Dysgenesis in Whom Pregnancy Was Terminated (*n* = 18)

CNS Diagnosis	Associated Findings	Gestational Age at Fetal MRI (wk/d)	Amniocentesis
Isolated inferior vermian hypoplasia	—	17/6	Normal
	Ambiguous genitalia	20/1	Smith-Lemli-Opitz syndrome
	—	20/2	Normal
	—	20/5	Normal
	—	21/0	Declined
	—	21/3	Normal
	—	21/4	Declined
Dandy-Walker malformation	HLHS, renal anomaly	18/5	Normal
	Short limbs, clenched fists	19/3	Normal
	TOF, echogenic kidneys	19/4	Declined
	—	20/5	Normal
	ACC, HLHS, ambiguous genitalia	21/4	Trisomy 13
Inferior vermian hypoplasia	ACC, renal dysplasia	20/0	Normal
	Ventriculomegaly	21/3	Normal
	Holoprosencephaly	22/3	Normal
Arachnoid cyst	ACC	22/4	Normal
Hypoplastic cerebellum	Micrognathia, echogenic bowel, hypertelorism	17/1	Normal
Rhombencephalosynapsis	Hydrocephalus	21/0	Normal

Note—Dash (—) indicates no associated findings. HLHS = hypoplastic left heart syndrome, TOF = tetralogy of Fallot, ACC = agenesis of the corpus callosum.

TABIE 4Cases with Discrepant Findings Between Fetal and Postnatal MRI studies (*n* = 16)

Fetal MRI Diagnosis (no. of cases)	Gestational Age at MRI (wk/d)	Postnatal MRI Diagnosis
Fetal MRI diagnoses excluded by postnatal MRI (6)		
Isolated inferior vermian hypoplasia (5)	18/3–22/0	
Mega cisterna magna (1)	22/0	
Cases with additional findings noted on postnatal MRI (10)		
Dandy-Walker malformation (1)	20/0	Dandy-Walker malformation, cerebral heterotopias
Dandy-Walker malformation, ACC (2)	19/3, 28/4	Dandy-Walker malformation, ACC, brainstem hypoplasia (both cases)
Dandy-Walker malformation, ACC, cerebral heterotopias (1)	23/1	Dandy-Walker malformation, ACC, ventriculomegaly, lissencephaly
Isolated inferior vermian hypoplasia (2)	17/4	Inferior vermian hypoplasia, cerebellar hypoplasia, cerebral heterotopias
	20/2	Inferior vermian hypoplasia, superior vermian dysplasia
Cerebellar hypoplasia, ACC (1)	18/2	Unilateral cerebellar hypoplasia, ACC, cerebral heterotopias
Cerebellar hypoplasia, inferior vermian hypoplasia (1)	22/0	Unilateral cerebellar hypoplasia, superior vermis dysplasia, contralateral cerebral underdevelopment
Arachnoid cyst (1)	23/3	Arachnoid cyst, cerebellar hypoplasia, cerebral heterotopias
Mega cisterna magna (1)	32/4	Posterior fossa hemorrhage

Note—ACC = agenesis of the corpus callosum.