Contribution of magnetic resonance imaging to the prenatal diagnosis of common congenital vascular anomalies

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Abstract

Background Screening ultrasound (US) has increased the detection of congenital vascular anomalies in utero. Complementary magnetic resonance imaging (MRI) may improve the diagnosis, but its real utility is still not well established.

Objectives We aimed to describe the imaging findings on prenatal US and MRI of the most frequent congenital vascular anomalies (lymphatic malformations and congenital hemangiomas) to assess the accuracy of prenatal US and MRI exams for diagnosis and to evaluate the relevance of the additional information obtained by complementary fetal MRI.

Materials and methods All confirmed postnatal congenital vascular anomalies detected in the last 10 years at 3 university hospitals were retrospectively identified. The prenatal diagnosis was compared with the final diagnosis for both methods and the clinical relevance of additional MRI information was evaluated. A second MRI in advanced pregnancy was performed in fetuses with lesions in a sensitive anatomical location and the clinical relevance of the additional information was evaluated.

Results Twenty-four cases were included in the study, 20 lymphatic malformations and 4 hemangiomas. MRI slightly improved the diagnosis of lymphatic malformation, 85% vs. 80% at US, especially for abdominal lesions. Both methods had a low identification rate (25%) for tumors. MRI performed late in five fetuses with lymphatic malformation allowed optimized management at birth.

Conclusion MRI improves the diagnosis of congenital lymphatic malformations whereas hemangiomas remain difficult to identify in utero. The main role of MRI is to provide high-defined anatomical data to guide management at birth.

Keywords Congenital hemangiomas · Fetus · Lymphatic malformations · Magnetic resonance imaging · Prenatal diagnosis · Ultrasound · Vascular anomalies

Introduction

After decades of confusing nomenclature, the International Society for the Study of Vascular Anomalies (ISSVA) adopted in 1996 the classification suggested by Mullicken and Glowacki in 1982 [1], which distinguishes between:

(1) Vascular malformations, which are developmental alterations of vascular channels. They are classified according to the predominant type of vessel in the malformation into arterial, venous, capillary, lymphatic or mixed [2, 3], and have a characteristic high- or low-flow pattern in the presence or absence of an arterial component [1, 2]. They may be part of an “overgrowth disorder” in a wide variety of
syndromes that combine a vascular malformations with a dysplastic, enlarged bone, limb or even a body-half [3, 4].

(2) Vascular tumors, which are formed by proliferative and hyperplastic endothelial lesions and typically have a high-flow arterial pattern. In contrast to the most common vascular tumor, benign infantile hemangioma, congenital hemangiomas are fully developed at birth. For a long time, vascular tumors were classified according to their postnatal clinical evolution into rapidly involuting congenital hemangiomas (RICH), which have a spontaneous and complete resolution without therapy in <14 months, and non-involuting congenital hemangiomas (NICH), which grow with the child, show no regression and may require therapy [2, 4, 5]. More recently, partially involuting hemangiomas (PICH) have been described as an intermediate type that initially behaves as a RICH lasting as a NICH [6].

The generalization of screening ultrasound (US) during pregnancy has increased the rate of detection of vascular anomalies in utero [7, 8]. An accurate prenatal diagnosis may help the medical team evaluate the extension and precise anatomical location of these lesions and improve the information provided to the parents, including predictions regarding prognosis during pregnancy, at birth and in the neonatal period [2, 9].

Prenatal magnetic resonance imaging (MRI) can be a complementary imaging method for fetal pathology, but its usefulness in patients with congenital vascular anomalies is not well known. The aims of this study are to describe the prenatal imaging findings on US and MRI of the two most frequently detected congenital vascular anomalies: lymphatic malformations and congenital hemangiomas. We compare the accuracy of both methods for diagnosis and evaluate the relevance of the additional information provided by complementary prenatal MRI for management.

Materials and methods

Inclusion criteria

A retrospective chart review of a 10-year period (January 2010 to December 2019) was conducted for all neonates with a confirmed congenital lymphatic malformation or hemangioma with available prenatal US and MRI exams.

The study was performed in three different hospitals, the University Hospitals of Lausanne (CHUV) and Genève (HUG) in Switzerland and the Necker Children’s Hospital in Paris (France), and was approved by the three institutional review boards and ethics committees. The clinical data of the patients included in the study were obtained from the Soarian program (Cerner, London, United Kingdom) at the Swiss hospitals/Mediweb program (Mediweb Solutions, Strasbourg, France) in France and the prenatal imaging exams (US and MRI) from the Archimède program (Archimède Solutions, Geneva, Switzerland) in Switzerland/Astraia program (Astraia Software gmbh, Ismaning, Germany) in France.

Classification of congenital vascular anomalies

Vascular anomalies were classified according to Mullicken and Glowacki’s classification accepted by the ISSVA [1] (Table 1).

Prenatal imaging studies

Screening US exams, including color Doppler studies, were usually performed between the 19th and 24th week of pregnancy by obstetricians with a wide experience in prenatal diagnosis with the following US systems: Voluson 730/E8/E10 Expert (GE Healthcare, Strasbourg, France) in Switzerland and France and/or Acuson Sequoia (Siemens Healthineers, Erlangen, Germany) in Switzerland.

Complementary MRI exams were performed using a phased-array multi-channel body coil on a 1.5-tesla (T) MR system with the machines and protocols described in Table 2. MRI exams were evaluated by experienced pediatric radiologists (L.A., P.S., A.-E.M., D.G. and S.H., all with more than 20 years of experience in radiology and more than 15 years of experience in pediatric radiology) who had access to the previous US images and reports.

Standard prenatal imaging reports included descriptions of the size, volume, anatomical location and organ of origin of the detected pathology. The morphology of the lesion — mostly solid or cystic, micro- or macrocystic, homogeneous or heterogeneous — its characteristics of echogenicity/signal intensity and its effects on the adjacent organs when performed were also included in the reports.

Review of prenatal studies

The reports of the prenatal imaging studies and their prospectively suggested diagnoses were obtained from the Radoffice (Medspazio, Geneva, Switzerland) and Soarian programs at the CHUV and Geneva and from Carestream (Philips, Horgen, Switzerland) at Necker. They were compared with the final diagnosis resulting from autopsy, anatomopathological exams, surgical reports, and/or postnatal clinical and imaging findings.

The rate of agreement between the prospectively suggested and final diagnoses for both US and MRI were established and the sensitivity for both methods compared. Finally, the clinical relevance of the additional information obtained at MRI and its consequences on management decisions were registered.
Additional value of MRI studies performed late in pregnancy

In some patients with lymphatic malformations, a second prenatal MRI was performed close to the due date to optimize management upon delivery, with identical protocol to that previously described. The changes in management secondary to the information provided in these late exams were registered and evaluated.

Results

The retrospective search identified 24 patients who fit the inclusion criteria, with 20 lymphatic malformations and 4 hemangiomas. The mortality rate was 16.7% (4/24 patients). Pregnancy was legally terminated in two fetuses with large lymphatic malformations and two neonates died at birth, one because of an extensive thoracic lymphatic malformation with bilateral pleural effusions and lung infiltration and one because of a rapidly growing hepatic hemangioma causing severe pulmonary hypoplasia.

Data concerning the gender of the fetuses, the anatomical location of the lesions, the main imaging findings, the prenatal suggested diagnosis, the final confirmed diagnosis and the outcome of the patients are described in Table 3 for lymphatic malformations and in Table 4 for congenital hemangiomas. These tables also show the correlation between the prenatal US and MRI and between the prenatal and the final diagnoses.

Congenital lymphatic malformations

Lymphatic malformation was suggested in the prenatal imaging of 17 patients and confirmed after birth for the last 3 cases (Table 3). Most of the lesions (14 cases; 70%) were located in the face and neck with occasional extension into the axilla and/or the thorax. The remaining six lesions were located at the axilla (one case), the thorax (two cases), or the abdomen and pelvis (three cases). Prenatal US achieved a correct diagnosis in 16 cases

| Tumors/ Malformations | Lymphatic malformations | Lymphatic-venous malformations |
|-----------------------|-------------------------|-------------------------------|
| RICH                  | Capillary malformations | Capillary-venous malformations |
| NICH                  | Lymphatic malformations | Lymphatic-venous malformations |
| PICH                  | Venous malformations    | Capillary-arterial-venous malformations |

Table 1 International Society for the Study of Vascular Anomalies classification of congenital vascular pathologies (adapted from [10])

| NICH non-involuting congenital hemangioma, PICH partially involuting hemangioma, RICH rapidly involuting congenital hemangioma

Additional value of MRI studies performed late in pregnancy

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Table 2 Magnetic resonance systems and protocols of fetal magnetic resonance imaging used in the three university hospitals

| Brand, Switzerland | Model | Sequences |
|---------------------|-------|-----------|
| Lausanne, Switzerland | Siemens Healthineers (Erlangen, Germany) | Magnetom Symphony Aera | In the 3 fetal planes: T2 half-Fourier acquisition single-shot turbo spin echo (HASTE) T2 true fast imaging with steady-state precession (FISP) T1-weighted volumetric interpolated breath-hold examination |
| Geneva, Switzerland | Siemens Healthineers | Avanto | T2 HASTE coronal T2 true FISP sagittal 2-dimensional T1-weighted spoiled incoherent gradient echo sequence axial |
| Paris, France | GE Healthcare (Waukesha, WI) | Optima MR450w | In the 3 fetal planes: Fast imaging employing steady-state acquisition (FIESTA) Single-shot fast spin echo 3-dimensional spoiled gradient echo pulse sequence |

| Springer |
| No. | Gender | Anatomical location | US diagnosis | MRI diagnosis | Concordance US/ MRI | Imaging findings and complications | Final diagnosis | Concordance pre-/postnatal | Final outcome |
|-----|--------|---------------------|--------------|--------------|---------------------|-----------------------------------|----------------|-----------------------------|---------------|
| 1   | M      | Left face superficial | LM           | LM           | Yes                 | Homogenous fluid content, No significant mass effect | Mixed LM       | Yes                         | Alive         |
| 2   | F      | Left face superficial | LM           | LM           | Yes                 | Homogenous fluid content, No significant mass effect | Mixed LM       | Yes                         | Alive         |
| 3   | M      | Left face + neck     | LM           | LM           | Yes                 | Homogenous fluid content, Tongue infiltration, tracheal compression, supra-aortic vessels encasement | Mixed LM       | Yes                         | Alive         |
| 4   | M      | Left face + neck     | LM           | LM           | Yes                 | Slightly heterogenous fluid content, No significant mass effect | Microcystic LM | Yes                         | Alive         |
| 5   | F      | Face + neck          | LM           | LM           | Yes                 | Homogenous fluid content, No significant mass effect | Mixed LM       | Yes                         | Alive         |
| 6   | M      | Face + neck          | LM           | LM           | Yes                 | Homogenous fluid content, No significant mass effect | Microcystic LM | Yes                         | Alive         |
| 7   | M      | Neck superficial     | LM           | LM           | Yes                 | Homogenous fluid content, No significant mass effect | Macrocystic LM | Yes                         | Unknown       |
| 8   | M      | Neck                 | LM           | LM           | Yes                 | Homogenous fluid content, No significant mass effect | Microcystic LM | Yes                         | Alive         |
| 9   | M      | Supra-hyoid neck     | LM           | LM           | Yes                 | Homogenous fluid content, Tongue infiltration, oropharynx displacement, jugulo-carotid vessels entrapment | Mixed LM       | Yes                         | Pregnancy termination |
| 10  | M      | Right neck superficial | LM           | LM           | Yes                 | Heterogenous fluid content, No significant mass effect | Macrocystic LM | Yes                         | Alive         |
| 11  | F      | Right neck + thorax  | LM           | LM           | Yes                 | Heterogenous fluid content, Infiltration of the thoracic wall and lung | Macrocystic LM | Yes                         | Alive         |
| 12  | M      | Left neck + thorax   | LM           | LM           | Yes                 | Heterogenous fluid content, Jugulo-carotid vessels encasement, Trachea displacement | Macrocystic LM | Yes                         | Alive         |
| 13  | M      | Right face + neck + thorax | LM | LM | Yes | Heterogenous fluid content, Tracheal compression and right lung invasion | Mixed LM       | Yes                         | Death at birth |
| 14  | M      | Right face + neck    | LM           | LM           | Yes                 | Homogenous fluid content, Oropharynx extension, Tongue invasion | Mixed LM       | Yes                         | Pregnancy termination |
| 15  | M      | Right axilla         | LM           | LM           | Yes                 | Heterogenous fluid content, No significant mass effect | Mixed LM       | Yes                         | Alive         |
| 16  | F      | Left thorax subcutaneous | LM         | LM           | Yes                 | Homogenous fluid content | Yes                         | Alive         |
### Table 3 (continued)

| N  | Gender | Anatomical location | US diagnosis | MRI diagnosis | Concordance US/MRI | Imaging findings and complications | Final diagnosis | Concordance pre-/postnatal | Final outcome |
|----|--------|---------------------|--------------|---------------|-------------------|-----------------------------------|----------------|-----------------------------|---------------|
| 17 | F      | Right thorax        | CPAM         | CPAM          | Yes               | - No significant mass effect      | Macrocystic LM | No                          | Alive         |
| 18 | M      | Abdomen intraperitoneal | Fetal peritonitis | LM            | No               | - Homogenous fluid content        | Macrocystic LM | Yes                         | Alive         |
| 19 | M      | Abdomen retroperitoneal | Teratoma     | Teratoma      | Yes               | - No significant mass effect      | Macrocystic LM | No                          | Alive         |
| 20 | M      | Pelvis presacral    | Teratoma     | Teratoma      | Yes               | - Homogenous fluid content        | Macrocystic LM | No                          | Alive         |

*CPAM* congenital pulmonary airway malformation, *LM* lymphatic malformation, *Mixed* macro- and microcystic lesion, *N* patient number

### Table 4

Data of the patients with congenital hemangioma included in this series: gender, anatomical location, prenatal suggested diagnosis, final diagnosis and clinical outcome

| N  | Gender | Anatomical location | US diagnosis | MRI diagnosis | Imaging findings and complications | Concordance US/MRI | Final diagnosis | Concordance pre-/postnatal | Final outcome |
|----|--------|---------------------|--------------|---------------|-----------------------------------|-------------------|----------------|-----------------------------|---------------|
| 21 | M      | Left scalp          | Teratoma     | Teratoma      | - Heterogenous                    | Yes               | Scalp hemangioma | No                          | Alive         |
| 22 | M      | Left hepatic lobe   | Mesenteric teratoma | Mesenteric teratoma | - No cardiovascular impact, - Heterogenous, - Hypervascular, - Enlarged left hepatic vein, - Cardiac failure | Yes               | Hepatic hemangioma | No                          | Alive         |
| 23 | F      | Left hepatic lobe   | Hepatic hamartoma | Hepatic hamartoma | - Very heterogenous, - Central cystic areas, - Cardiac failure, - Hydrops fetalis, - Bilateral lung hypoplasia | Yes               | Hepatic hemangioma | No                          | Death at birth |
| 24 | M      | Left hepatic lobe   | Hepatic hemangioma | Hepatic hemangioma | - Heterogenous, - Hypervascular, - Enlarged left hepatic vein, - Cardiac failure | Yes               | Hepatic hemangioma | Yes                         | Alive         |

*N* patient number
with the following anatomical locations: cervical and/or facial (11 cases), axillary (1 case), cervicothoracic (3 cases) or thoracic (1 case). Prenatal MRI identified 17 cases (85%), the 16 first previously described at US and 1 additional abdominal lesion. In three fetuses with a postnatally confirmed lymphatic malformation, prenatal US and MRI suggested a teratoma for two abdominal lesions (Fig. 1) and a congenital pulmonary airway malformation (CPAM) for a thoracic lesion (Table 3). The only discrepancy between the suggested diagnosis on US and MRI was an extensive mesenteric lymphatic malformation, which was correctly diagnosed on MRI whereas US suggested a peritonitis after intestinal perforation (Fig. 2).

**Additional value of MRI studies performed late in pregnancy**

In five fetuses with prenatally diagnosed lymphatic malformations, a second MRI exam was performed during advanced pregnancy (33–36 weeks). Indication for these late studies were an anatomical location close to the upper respiratory airways (five cases) and/or a voluminous lesion (two cases) (Fig. 3). These exams were performed for management at birth, including decisions about the type of delivery and the need for extracorporeal intrapartum treatment (EXIT procedure) [11]. Table 5 includes the additional information provided by these late performed MRIs and its influence on patient management.

**Congenital hemangiomas**

The anatomical location of the four confirmed congenital hemangiomas included in this series were the scalp (one case) and the liver (three cases) (Figs. 4 and 5). The prenatal accuracy of diagnosis was very low and showed no difference between the two imaging methods, with only 1 of 4 (25%) confirmed hemangiomas correctly identified in utero (Table 4).

**Discussion**

The ISSVA acceptance of Mullicken and Glowacki’s classification for congenital vascular anomalies [1] has contributed considerably to a better understanding of these pathologies while the generalization of screening US has increased their in utero detection rate. Impressive technical advances and an increased availability of MRI have led to an extension of this
method for diagnosing prenatal pathology. However, data concerning its real contribution for congenital vascular anomalies remain limited [12–15].

**Congenital lymphatic malformations**

Lymphatic malformation is the most often detected vascular malformation in the fetus and the most widely described in the literature [4, 15–18]. The reported incidence is 1,200–6,000 cases in live births, with a slight male predominance [16]. They can occur in any location, but are much more often located in the neck (75%) and the axilla (20%) than in the abdomen (2%), limbs (2%) and mediastinum (1%) [4, 15, 17]. They are usually classified as microcystic (cysts <2 cm in size), macrocystic (>2 cm) and mixed lesions. Macrocytic lesions are usually more voluminous and therefore easier to detect in utero than microcystic ones [17]. At US, these malformations appear as multiseptated cystic lesions, lacking solid components, vascularity and calcifications. However, a recent review has described a greater imaging variability than previously reported, including mixed cystic/solid lesions and occasional calcifications [17].

Our series evaluated 20 confirmed lymphatic malformations and obtained a high accuracy in the detection and correct identification of these lesions on both prenatal US and MRI, 80% and 85%, respectively. All our cervicofacial and axillary located malformations were correctly identified on both methods and compared to previously published series [4, 15, 17] we had a higher percentage of rare anatomical locations, including the thorax (2 cases; 10%) and the abdomen and/or pelvis (3 cases; 15%). We observed a significant discordance between the prenatal suggested diagnosis and the final diagnosis in these uncommon locations. US identified only one of two thoracic and none of three abdominal

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Fig. 2 A male fetus at 35 weeks’ gestation with extensive mesenteric malformation and postnatal surgery at 4 months old (N18, Table 3). a–d Axial US (a) and T2-weighted MR (b) images (repetition time [TR]/echo time [TE] 1,200/90 ms) at the same level and coronal T2- (c) (TR/TE 1,200/90 ms) and T1-weighted (d) (TR/TE 3.29/1.29 ms) MR images show the extensive mesenteric macrocystic lymphatic malformation with homogenous liquid echogenicity and signal intensity and multiple internal septations. The lesion displaces the fetal intraperitoneal organs but shows no significant complication. The fetal colon, surrounded by the lymphatic malformation, is easily identifiable by its meconium filling, hypointense on T2- and hyperintense on T1-weighted images (arrows). The lesion was correctly identified only at MRI. e Postnatal image during the surgical procedure shows the voluminous lesion surrounding the colon.
malformations, whereas MRI identified one thoracic and one abdominal lesion. For the two remaining unidentified cases of abdominal lesions, both methods suggested the teratoma, probably influenced by the presacral location of one of these lesions, typical for this common fetal tumor (Fig. 1).

Atypical imaging findings were the second explanation for the discordance between the prenatal and the final diagnosis. As previously reported, the differentiation between a lymphatic malformation and a fetal teratoma can be extremely difficult in the presence of calcifications, hemorrhage or of mixed cystic and solid lesions [17]. Indeed, one of the misinterpreted mesenteric lymphatic malformations showed an extensive hemorrhage with marked heterogeneity in utero.

Additional information provided by MRI

MRI is less affected than US by fetal position, oligohydramnios, maternal obesity and fetal bone superposition and can more precisely determine the extent of a lesion and its relationship to the adjacent structures [16, 18, 19]. In our series, the information provided by complementary MRI improved the echographic suggested diagnosis and changed the management of 2 (10%) patients. In a thoracic lymphatic malformation, MRI showed an extension toward the mediastinum and into the lungs in addition to voluminous pleural effusions already detected on US, which led to a pregnancy continuation with only comfort care at birth. In a second case, MRI correctly identified a voluminous mesenteric lymphatic malformation and excluded relevant complications, which led to the decision to continue the pregnancy (Fig. 2).

MRI studies performed late in pregnancy

In cervically located lymphatic malformations, MRI can document the extension toward the mediastinum and evaluate the proximity to the brachial plexus [16, 18] or the degree of compression of the airways [11, 18, 19], information difficult to obtain by US alone [13]. A second MRI exam at advanced pregnancy performed in five lymphatic malformations located in a sensitive anatomical location provided accurate anatomical information and enabled decisions about the type of delivery (Table 5), including the successful performance of an EXIT procedure in a fetus showing increasing tracheal compression (Fig. 3).

Congenital hemangiomas

Congenital hemangiomas are already fully developed at birth [20]. RICH and NICH types have an almost equal gender distribution, are usually solitary lesions and have a predilection for the skin, mostly in the head or limbs. However, the reported imaging findings of these lesions are still limited,

| N  | Location                  | Time of MRI (weeks of pregnancy) | Additional information at 2nd MRI                                                                 | Management                                      |
|----|---------------------------|----------------------------------|---------------------------------------------------------------------------------------------------|------------------------------------------------|
| 3  | Left face + neck          | 32/35                            | - Major volume augmentation without hemorrhage                                                    | EXIT procedure                                 |
|    |                           |                                  | - Oropharynx distortion                                                                         | Emergency cesarean at 36 weeks of pregnancy    |
|    |                           |                                  | - Increasing tracheal compression                                                                |                                                |
| 5  | Face + neck               | 26/34                            | - No significant tracheal displacement                                                             | Normal birth                                   |
|    |                           |                                  | - No hemorrhage                                                                                  |                                                |
| 11 | Right neck + thorax       | 25/36                            | - Major volume augmentation due to hemorrhage                                                     | No need of EXIT                                |
|    |                           |                                  | - Slight lateral tracheal displacement                                                            | Elective cesarean at 37 weeks of pregnancy     |
|    |                           |                                  | - Extreme arm abduction                                                                          | Immediate intubation                           |
| 12 | Left neck + thorax        | 22/34                            | - Mediastinal extension                                                                          | No need of EXIT                                |
|    |                           |                                  | - Slight tracheal displacement                                                                    | Elective cesarean at 39 weeks of pregnancy     |
|    |                           |                                  | - No hemorrhage                                                                                  | Immediate intubation                           |
| 18 | Abdomen intraperitoneal   | 26/35                            | - Major volume augmentation without hemorrhage                                                    | No need of EXIT                                |
|    |                           |                                  | - Increasing abdominal distention                                                                  | Elective cesarean at 38 weeks of pregnancy     |
|    |                           |                                  | - No intestinal obstruction                                                                      | Surgical excision at 2 months of age            |

EXIT ex utero intrapartum treatment, N patient number

Table 5 Additional information provided by late pregnancy MRI exams in patients with lymphatic malformations
with descriptions of isolated cases, concerning mainly RICH-type lesions in typical locations [5, 12, 21].

Hemangiomas are the most frequently detected liver tumors in fetuses and neonates, concerning more than 60% of all congenital hepatic lesions [22]. They are often heterogeneous on US and may contain identifiable calcifications. On MRI, they also are frequently heterogeneous, with foci of hyperintensity on T1-weighted images and hypointensity on T2-weighted images and intratumoral flow void that represents tubular vascular structures. In a series of 16 children, Franchi-Abella et al. [23] reported well-defined, heterogeneous, hypoechogenic lesions compared to the normal liver on US with intratumoral abnormal vessels and enlarged hepatic arteries and/or veins. On MRI, they showed a high signal intensity on T2-weighted images and a low signal intensity on T1-weighted images when compared to the normal liver, with intraliesional signal flow voids. Jiao-Ling et al. [24] described well-defined masses on US exams in a series of six congenital hepatic hemangiomas that were mostly hypoechogenic compared to the normal liver with significant rates of heterogeneity, necrosis, cystic cavities and calcifications. Color Doppler showed enlarged hepatic arteries and tortuous, dilated veins. The lesions were hypointense on T1-weighted MR images and hyperintense on T2-weighted images compared to the normal liver.

In our series, the sensitivity of prenatal US and MRI for congenital hemangiomas was only 25%, and showed no difference between both prenatal methods. Only one of three hepatic hemangiomas was identified in utero. (Table 4). Strangely, the three hepatic tumors were exophytic, growing from the edge of the left lobe. Two of them had almost identical imaging findings and were hypoechogenic, heterogeneous on US and mostly hypointense on T2-weighted MR images compared to the normal liver with clearly visible flow voids and an extremely enlarged left hepatic vein (Fig. 4). These two lesions were classified as RICH tumors according to the postnatal evolution. In contrast, the third hemangioma was a rapidly growing, heterogeneous lesion with extensive avascular areas on US Doppler and no intratumoral flow voids identifiable on MRI. Autopsy after emergency cesarean and death at birth revealed a huge hemangioma with extensive areas of hemorrhage, thrombosis and necrosis (Fig. 5).

**Additional information provided by MRI**

Complementary MRI influenced the management in a rapidly growing hepatic congenital hemangioma while revealing significant bilateral pulmonary hypoplasia. Emergency cesarean was performed, but the patient died at birth from combined cardiorespiratory insufficiency.

Franchi-Abella et al. [23] suggested that fetal US should remain the standard diagnostic method for hepatic hemangiomas and proposed that MRI should only be performed when the diagnosis is unclear or the lesion is not well-delimited. In opposite, Jiao-Ling et al. [24] and our own results show that voluminous hepatic hemangiomas often have a great imaging variability that could explain the low rates of echographic diagnosis in utero. Although our prenatal rate remained low after MRI, we still believe these exams helped evaluate the effects of the lesions on the fetus.

This article increases the limited available information regarding the prenatal imaging findings of the most frequently
**Fig. 4** A male fetus at 33 weeks’ gestation (N22) with hepatic hemangioma and postnatal images at 4 days old. 

a. Transverse US image shows a voluminous, solid appearing, heterogeneous left hepatic mass. Arrows indicate the borders of the lesion. S: spleen, St: stomach. 

b–d. Axial Doppler US image (b), coronal T2-weighted (c) (repetition time [TR]/echo time [TE] 1,000/92 ms) image at the same level as well as axial T2-weighted (d) MR image (TR/TE 6.60/3.30 ms) show the inhomogeneous, exophytic growing lesion arising from the left hepatic lobe (H in b, arrowheads in c, d). Note the numerous tubular forming flow voids indicating vascular structures (arrow in d) and the extreme enlarged left hepatic vein (arrow in b). 

e. Postnatal T1-weighted MR image (TR/TE 3.61/1.61 ms) after contrast shows the marked early peripheral enhancement of the lesion at the arterial phase and confirms the enlarged left hepatic vein (arrow). After significant regression, the tumor was resected when the boy was 18 months old.

**Fig. 5** A male fetus at 27 weeks’ gestation with hepatic hemangioma (N23). 

a–b. Coronal (a) and left sagittal (b) T2-weighted MR images (repetition time/echo time 1,530/86 ms) show a huge, heterogeneous left abdominal mass arising from the left hepatic lobe. Note the absence of tubular intralesional structures. Arrowheads show the borders of the lesion. The tumor causes bilateral elevation of the hemidiaphragms and severe secondary lung hypoplasia, fetal cardiomegaly and marked hydrops fetalis. The patient died at birth after an emergency cesarean at the 27th week of pregnancy. 

c. Macroscopic view of the fetal liver confirms the hepatic origin of this tumor and reveals central cavities and extensive necrosis.
detected congenital vascular anomalies and reveals the main difficulties in their prenatal diagnosis. However, it has some limitations, including its retrospective character and a reduced number of patients despite the participation of three university hospitals. Therefore, these results should be confirmed by more extensive and prospective studies.

Conclusion

Macrocytic lymphatic malformations and rapidly involuting congenital vascular anomalies are the most commonly detected congenital vascular anomalies in utero. A rare anatomical location and atypical imaging findings, such as hemorrhage or necrosis, complicate their prenatal diagnosis. The main role of complementary MRI is probably not to improve the diagnosis but to provide high-defined anatomical data to guide the management and anticipate complications at birth.

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Declarations

Conflicts of interest

None

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