Prenatal Diagnosis of Vascular Anomalies

By Jennifer J. Marler, Steven J. Fishman, Joseph Upton, Patricia E. Burrows, Harriet J. Paltiel, Russell W. Jennings, and John B. Mulliken
Boston, Massachusetts

Background/Purpose: Vascular anomalies are diagnosed prenatally with increasing frequency. The authors reviewed a group of children treated at their center who had an abnormal prenatal diagnosis to determine (1) fetal age at which the vascular anomaly was detected, (2) general diagnostic accuracy, and (3) impact on ante- and postnatal care. Their findings are compared with reported cases and series. The authors clarify appropriate terminology and underscore the need for interdisciplinary participation of specialists in the field of vascular anomalies.

Methods: Patients referred during prenatal life and children with a history of abnormal antenatal findings seen at our vascular anomalies center during a 1-year period (September 1999 through August 2000) were included in this study. The fetal age at diagnosis, pre- and postnatal diagnoses, antenatal course, and neonatal outcome were obtained from the parents, through chart reviews, and through telephone interviews with the treating obstetricians.

Results: Twenty-nine patients with vascular anomalies were identified: 17 had a correct prenatal diagnosis, and 12 had an incorrect diagnosis, an overall diagnostic accuracy of 59%. Capillary-lymphatic-venous malformations (CLVM) most often were correctly diagnosed (67%), followed by lymphatic malformation (LM, 62%) and hemangioma (59%). In the infants who received correct diagnoses in utero, there were no fetal deaths and there was no neonatal morbidity. Maternal steroids were administered for a fetus with an intrahepatic hemangioma and deteriorating cardiac function, with subsequent stabilization and successful delivery of a healthy neonate. Among infants with incorrect diagnoses, there was 1 postnatal death, 1 case of erroneous gender assignment, one case of unnecessary fetal surgical intervention, 1 unnecessary neonatal laparotomy, and 1 delay in diagnosis of a malignancy. Cesarean section was done for 65% of correctly diagnosed cases, (including 2 ex utero intrapartum [Exit] procedures) and for 33% of incorrectly diagnosed cases. Most diagnoses were made during the mid- to late second trimester and third trimester; only 4 cases (14%) were detected before 20 weeks.

Conclusions: In this series, accurate diagnosis optimized antenatal care by providing an opportunity for planning deliveries, for pharmacologic fetal intervention in 1 case, and for appropriate parental counselling. Inaccurate diagnosis was associated with significantly increased morbidity and mortality. Finally, the intrauterine diagnosis of LM should be distinguished from posterior nuchal translucency, an obstetric term applied to fetal lymphatic abnormalities detected in the first and second trimesters that do not manifest as postnatal LM.
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INDEX WORDS: Vascular anomalies, hemangioma, lymphatic malformation, capillary lymphaticovenous malformation, arteriovenous malformation, Klippel-Trenaunay syndrome, prenatal ultrasonography, prenatal diagnosis, fetal intervention, posterior nuchal translucency.

VASCULAR ANOMALIES are encompassed in 2 major categories: (1) vascular tumors, which are proliferative endothelial lesions, such as hemangiomas, and (2) vascular malformations, which are developmental aberrations of hematic or lymphatic vessels.[1] At our vascular anomalies center, we have received several referrals for affected fetuses. In addition, a number of our patients were documented to have prenatal pathology, some of whom had an inaccurate prenatal diagnosis. There now are numerous case reports of vascular anomalies detected in utero (Table 1). However, case reports published to date have been sporadic, often including small numbers of patients. In addition, there is a tendency for incorrectly diagnosed cases to go unpublished. This study was designed to determine the accuracy of prenatal ultrasonography in detecting vascular anomalies. Specificity and sensitivity of this modality for diagnosing vascular anomalies could not be assessed because we focused on a group of patients with these disorders rather than on a mixed population of children with and without disease.

MATERIALS AND METHODS

Patients presenting to our vascular anomalies center during a 1-year period (September 1999 through August 2000) who were referred

From the Departments of Surgery and Radiology, the Vascular Anomalies Center, and the Advanced Fetal Care Center, Children's Hospital, and Harvard Medical School, Boston, MA.

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Address reprint requests to Steven J. Fishman, MD, Vascular Anomalies Center, Department of Surgery, Children's Hospital, 300 Longwood Ave, Boston, MA 02115.

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Table 1. Data for Reported Cases of Prenatally Diagnosed Vascular Anomalies

Study	Fetal Age at Diagnosis (wk)	Diagnosis	Associated Findings	Type of Delivery	Ante-Neonatal Mortality Rate
Sheu et al[18]	NA	Hemangiomas—multiple	Hydrops	NA	Neonatal death
Nakamoto et al[19]	31	Hemangioma—hepatic	Polyhydramnios CHF, hydrops fetalis	NA	Intrauterine death
Abuhamad et al[20]	16	Hemangioma—hepatic	Abdominal distension	Termination at 22 weeks	
Sepulveda et al[21]	33	Hemangioma—hepatic	None	VD	
Walton et al[16]	21	Hemangioma—hepatic	Mild hydrops, cardiomegaly, truncal edema	CS	
Dreyfus et al[22]	36	Hemangioma—hepatic	None	CS	
Morris et al[6]	17	Hemangioma—hepatic	Cardiomegaly	CS	
Pennell and Baltarowich[23]	30	Hemangioma—scalp	None	CS	
Sherer et al[24]	24	Hemangioma—scalp	None	VD	
Mitchell[25]	22	Hemangioma—scalp	None	CS	
Schwartz et al[26]	30	Hemangioma—cervicofacial	Polyhydramnios	CS/Exit	
Meizner et al[27]	32	Hemangioma—facial	Polyhydramnios	NA	Intrauterine death
Eckstein et al[28]	34	Hemangioma—atrial	None	VD	
Tseng et al[29]	28	Hemangioma—atrial	Twin pregnancy pericardial effusion	CS	
Maynor et al[30]	38	Hemangioma—abdominal	None	VD	
Goncalves et al[31]	36	Hemangioma—thigh	None	CS	
Sheiner et al[32]	Third trimester	Hemangioma—thigh	Pericardial effusion	VD	
Treadwell et al[33]	36	Hemangioma—thigh and abdomen	None	CS	
Suma et al[34]	28	Hemangioma—thigh	None	CS	
Benacerraf and Frigoletto[14]	30-38	LM, 5 cases—3 cervical, 1 flank, 1 intraabdominal	Hydrops in one fetus with cervical LM	NA	Death of hydropic infant at birth secondary to inability to ventilate
Giacolone et al[35]	25	LM—cervical mediastinal, and abdominal	None	Termination	
Goldstein et al[36]	30-32	LM, 2 cases—1 axillary, 1 abdominal wall	None	CS	
			None	VD	
McCoy et al[37]	18	LM—axillary, bilateral	None	CS	
Thomas[38]	27	LM—cervical	None	CS	
Hoffman-Tretin et al[39]	36	LM—axillary	None	CS	
Zalel et al[40]	29	LM—mediastinal	None	VD	
Salvador et al[41]	31	LM—intraabdominal	None	CS	
Malnofski et al[42]	36	LM—retroperitoneal	None	CS	
Katz et al[43]	28	LM—lower extremity and flank	None	VD	
Hatjis et al[44]	34	CLVM	Polyhydramnios	VD	
Katz et al[43]	33	CLVM	Polyhydramnios	VD	Neonatal death

NOTE. This literature review does not include patients reported to have "eponymous" vascular anomalies where correct classification of the disorder could not be ascertained. For example, the incorrect term "Klippel-Trenaunay-Weber" precludes determination of whether a patient has Klippel-Trenaunay syndrome (CLVM) or Parkes Weber syndrome (CAVM). Nor did we include case series in which a diagnosis of posterior nuchal translucency could not be distinguished clearly from LM. We also excluded reported cases of "cavernous hemangioma" because this term does not differentiate between true hemangioma and venous malformation; it has been applied to both.

Abbreviations: CS, cesarean section; VD, vaginal delivery; NA, not available; Dx, diagnosis.

prenatally or whose parents reported a prenatally diagnosed abnormality were included in this study. This study was approved by the Committee on Clinical Investigation of Children's Hospital, Boston. Specific information obtained included prenatal and postnatal diagnosis, fetal age at diagnosis, associated anomalies (such as polyhydramnios or fetal hydrops), mode of delivery (vaginal, cesarean, or cesarean with exit), and morbidity or mortality during the first month of life. These data were collected from interviews with parents, review of pre- and postnatal charts, and discussions with the involved obstetrician.

There were multiple obstetricians and radiologists with varying levels of expertise involved in prenatal diagnosis. Postnatal diagnoses were assigned by our team of vascular anomaly physicians who reached a consensus after patient examination (where possible) or review of medical history, clinical photographs, radiographic studies, and laboratory studies. Our aim was to compare diagnoses during fetal life with those postnatally. The specific criteria that each ultrasonographer used to establish each prenatal diagnosis is outside the scope of this report.

Table 2. Data for Patients in Current Series

Patient No.	Sex	Fetal Age at Diagnosis (wk)	Postnatal Diagnosis	Prenatal Diagnosis	Associated Findings	Delivery	Ante- and Neonatal Mortality And Morbidity
1	M	16	Hemangioma—scalp	Hemangioma	None	CS	Healthy neonate
2	M	18	Hemangioma—scalp	Hemangioma	None	VD	Healthy neonate
3	M	24	Hemangioma—hepatic	Hemangioma	Cardiomegaly	CS	Healthy neonate
4	F	35	Hemangioma—hepatic and intracranial	Hemangioma	Polyhydramnios	CS	Healthy neonate
5	M	30	Hemangioma—hepatic	Hemangioma	Polyhydramnios	CS	Healthy neonate
6	F	37	Hemangioma—atrial	Cardiac teratoma	Polyhydramnios	CS	Healthy neonate
7	M	32	Hemangioma—leg and perineum	Large foot, female sex	None	VD	CHF, multiple Resections and amputation.
8	M	32	Neuroblastoma	Hemangioma—hepatic	None	CS	Healthy neonate
9	F	34	LM—anterior cervical	LM	None	CS/EXIT	Intubated during EXIT Tracheostomy in neonatal period.
10	F	32	LM—anterior cervical	LM	None	CS	Healthy neonate
11	F	28	LM—axillary, arm	LM	None	CS	Healthy neonate
12	M	24	LM—cervical	LM	None	VD	Healthy neonate
13	F	34	LM—intraabdominal	LM	None	VD	Healthy neonate
14	F	24	LM—anterior cervical	LM	None	CS	Healthy neonate
15	F	24	LM—thoracic	LM	None	CS	Healthy neonate
16	F	32	LM—axillary	LM	None	VD	Healthy neonate
17	F	30	LM—axillary, arm	LM	None	CS	Healthy neonate
18	M	16	LM—anterolat cervical	LM versus teratoma	None	CS/EXIT	Healthy neonate
19	F	20	LM—hand	Amniotic band syndrome	None	CS	Fetal surgery followed by premature delivery. Resection during infancy.
20	M	32	LM—cervicofacial	Encephalocele	None	VD	Healthy neonate
21	M	28	LM—sacrococcygeal	Teratoma	None	VD	Healthy neonate
22	F	37	LM—sacrococcygeal	Teratoma	None	VD	Healthy neonate
23	F	33	LM—axillary	Ectopia cordis	None	VD	Healthy neonate
24	M	17	LM—intraabdominal	Intrauterine bowel perforation	None	VD	Laparotomy during first week of life
25	F	33	AVM—leg	Hydropic female	Fetal ascites, hydrops	CS	CHF, neonatal embolization neonatal death
26	M	32	VM—blue rubber bleb nervus syndrome	Intraabdominal teratoma	None	VD	Healthy neonate
27	F	35	CLVM—trunk, leg	Abdominal mass	None	VD	Healthy neonate
28	F	35	CLVM—trunk, leg	CLVM	Polyhydramnios, pericard effusion, cystic kidney	VD	Healthy neonate
29	F	28	CLVM—leg	CLVM	None	VD	Healthy neonate

Abbreviations: CS, cesarean section; VD, vaginal delivery.

RESULTS

The results are summarized in Tables 2, 3, and 4 with examples presented in Figs 1 and 2. There were 29 patients, including 16 with lymphatic malformation (LM), 7 with hemangioma, 3 with capillary-lymphatic-venous malformation (CLVM), 1 with arteriovenous malformation (AVM), 1 with "Blue Rubber Nevus Syndrome" (BRBNS, a form of venous malformation [VM]) and 1 with neuroblastoma (diagnosed antenatally as a hepatic hemangioma).

Seventeen patients (59%) had a correct diagnosis, whereas 12 (41%) had an incorrect diagnosis. In the correct diagnosis group, there were 5 hemangiomas, 10 LMs and 2 CLVMs. Cesarean was the most common mode of delivery (11 cases, 64%); 2 of these were ex utero intrapartum (EXIT) procedures, both done for LMs

Table 3. Percentages of Correct and Incorrect Prenatal Diagnoses for All Cases and for Each Type of Vascular Anomaly

Vascular Anomaly	No.	Percent Correct Prenatal Diagnoses	Percent Incorrect Prenatal Diagnoses
All cases	29	59	41
Hemangioma	7	57	43
LM	16	62	38
CLVM	3	67	33
AVM	1	0	100
VM/BRBNS	1	0	100
Other diagnoses	1	0	100

threatening the airway. One patient required intubation with deferred tracheostomy, and another required immediate tracheostomy. Vaginal delivery accounted for 35% (6 cases). There were no deaths nor neonatal morbidity in this group.

The incorrect diagnosis group (n = 12) included 2 hemangiomas, 6 LMs, 1 CLVM, 1 AVM, and 1 BRBNS. There was 1 neonatal death—an infant with fetal congestive heart failure and hydrops diagnosed prenatally. Embolization failed to control the high flow through a massive AVM of the pelvis and lower extremity, which became obvious when the infant was delivered.

There was notable morbidity in the incorrect diagnosis hemangioma subgroup. One infant with a hepatic hemangioma diagnosed antenatally was found, on postnatal imaging and biopsy, to have neuroblastoma. A second infant, a boy with a perineal, pelvic, and extremity hemangioma, received an incorrect gender assignment, presumably secondary to scrotal swelling mimicking the appearance of female labia. Although his right lower extremity was detected to be larger prenatally, there was no documentation of increased blood flow through a massive hemangioma with arteriovenous fistulous components. Although there was no evidence of cardiac failure in utero, this infant became critically ill with high output cardiac failure, resulting in a long distance emergent transfer to our institution. Postnatally, this infant required 20 resections for nonviable extremity and perineal tissue (finally resulting in a high thigh amputation) and a number of embolizations to control congestive heart failure. A third infant was felt to have a cardiac teratoma that was found to be a right atrial hemangioma on surgical exploration at 3 days of life.

Additional morbidity was seen among incorrectly diagnosed LMs. One patient referred to our center postnatally with an LM of the hand had a prenatal diagnosis of "amniotic band syndrome." Fetal surgery was performed with laser release of the suspected amniotic band over the volar wrist. This was followed by premature delivery of the infant. When the infant was several months old, a dorsal debulking of the malformation was done at our institution. Postoperatively, there was profound venous engorgement of the hand that required application of leeches. This complication was ascribed to impaired venous drainage of the volar hand because of deep scar secondary to fetal intervention (Fig 1). A second infant with LM was diagnosed as having intrauterine bowel perforation; surgical exploration on the first day of life revealed intraabdominal LM.

In the incorrect diagnosis group, vaginal deliveries were most common (8 cases, 67%); cesarean sections were done in the remaining cases (4 cases, 33%) with no EXIT procedures.

DISCUSSION

On the basis of cellular kinetics and clinical behavior, there are 2 major categories of vascular anomalies—tumors and malformations.[1,2] The most common vascular tumor is hemangioma of infancy. Approximately 30% of hemangiomas are nascent at birth, presenting as a premonitory cutaneous mark, whereas two thirds of hemangiomas appear postnatally (typically around 2 weeks of age).[1] There is an uncommon variant termed *congenital hemangioma* that evolves during fetal life and manifests fully developed at birth. This subset has the greatest potential to be detected prenatally. Congenital hemangiomas are subdivided further according to their postnatal behavior. Most lesions involute more rapidly than common postnatal infantile hemangiomas ("rapidly-involuting congenital hemangiomas"),[3] whereas others

Table 4. Mode of Delivery and Neonatal Morbidity and Mortality for All Cases and for Correctly and Incorrectly Diagnosed Cases

	Vaginal Delivery	Cesarean	Cesarean + EXIT Procedure	Neonatal Mortality Rate	Neonatal Morbidity Resulting From Incorrect Diagnosis
All cases (n = 29)	14 (48%)	13 (51%)	2 (<1%)	1 (0.3%)	
Correctly diagnosed (n = 17)	6 (35%)	9 (53%)	2 (12%)	0	None noted
Incorrectly diagnosed (n = 12)	8 (67%)	4 (33%)	0	1 (0.8%)	One incorrect gender assignment. One missed malignancy. One unnecessary fetal surgery. One unnecessary neonatal laparotomy.

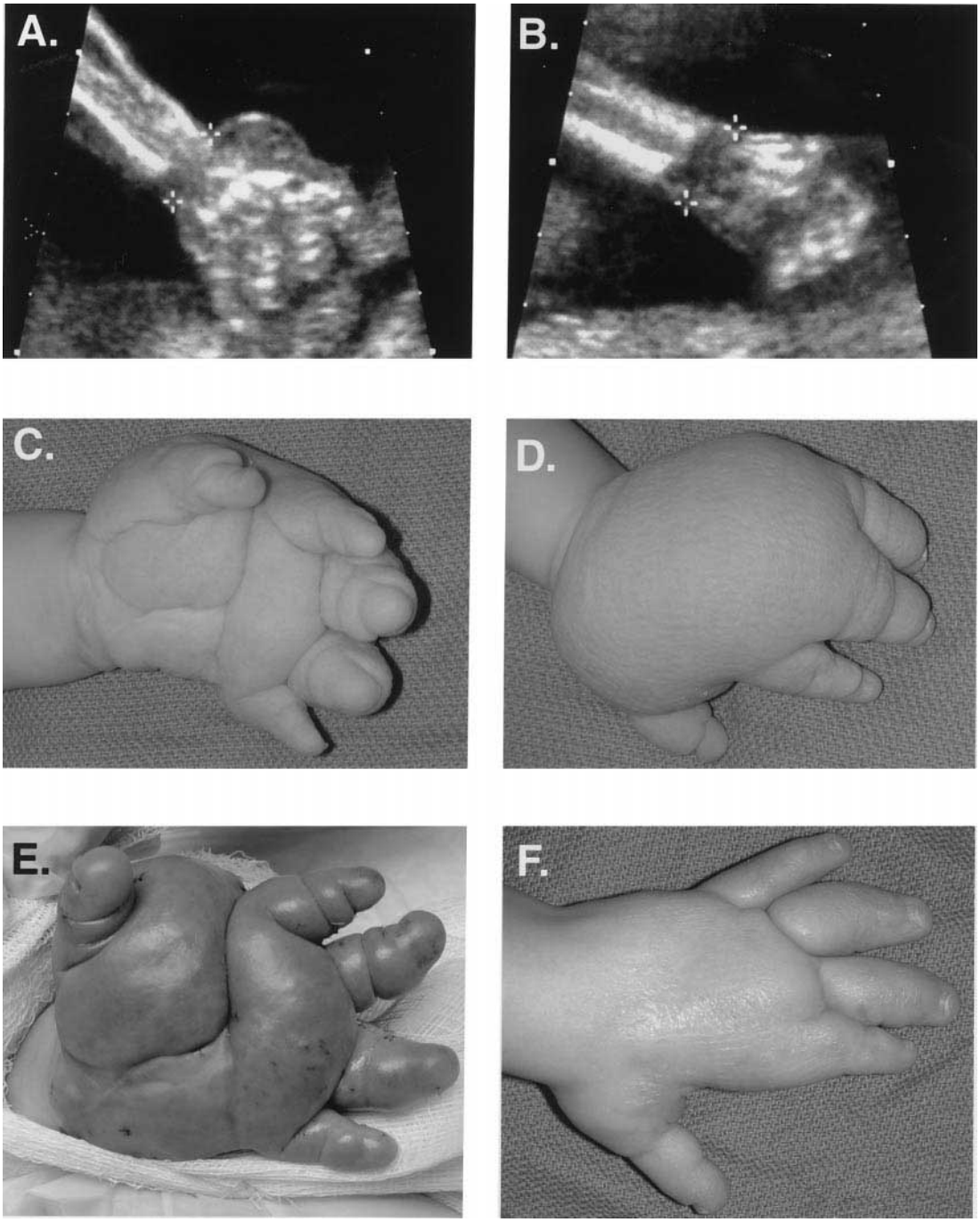

Fig 1. Complication secondary to a lymphatic malformation of the hand prenatally diagnosed as "amniotic band syndrome." (A) Fetal ultrasonogram of left hand at 22 weeks' gestation, compared with right hand (B). Hand at 4 months of age—volar (C) and dorsal (D) views. Scar from laser surgery of the fetus can be appreciated on volar surface of the wrist in (C). (E) Postoperative venous congestion after dorsal debulking. (F) Appearance of hand 3 months after second staged resection.

do not regress and persist into late childhood ("noninvoluting congenital hemangiomas").[4]

In our series, 4 of 7 hemangiomas were diagnosed correctly by sonography. Two of these were congenital hemangioma of the scalp, discovered early during the second trimester. Although they showed signs of regression during the first few weeks of life, they were excised in infancy. Bleeding and ulceration at a site at which the anticipated scar from ulceration will be more pronounced than that from excision is one of few indications for early

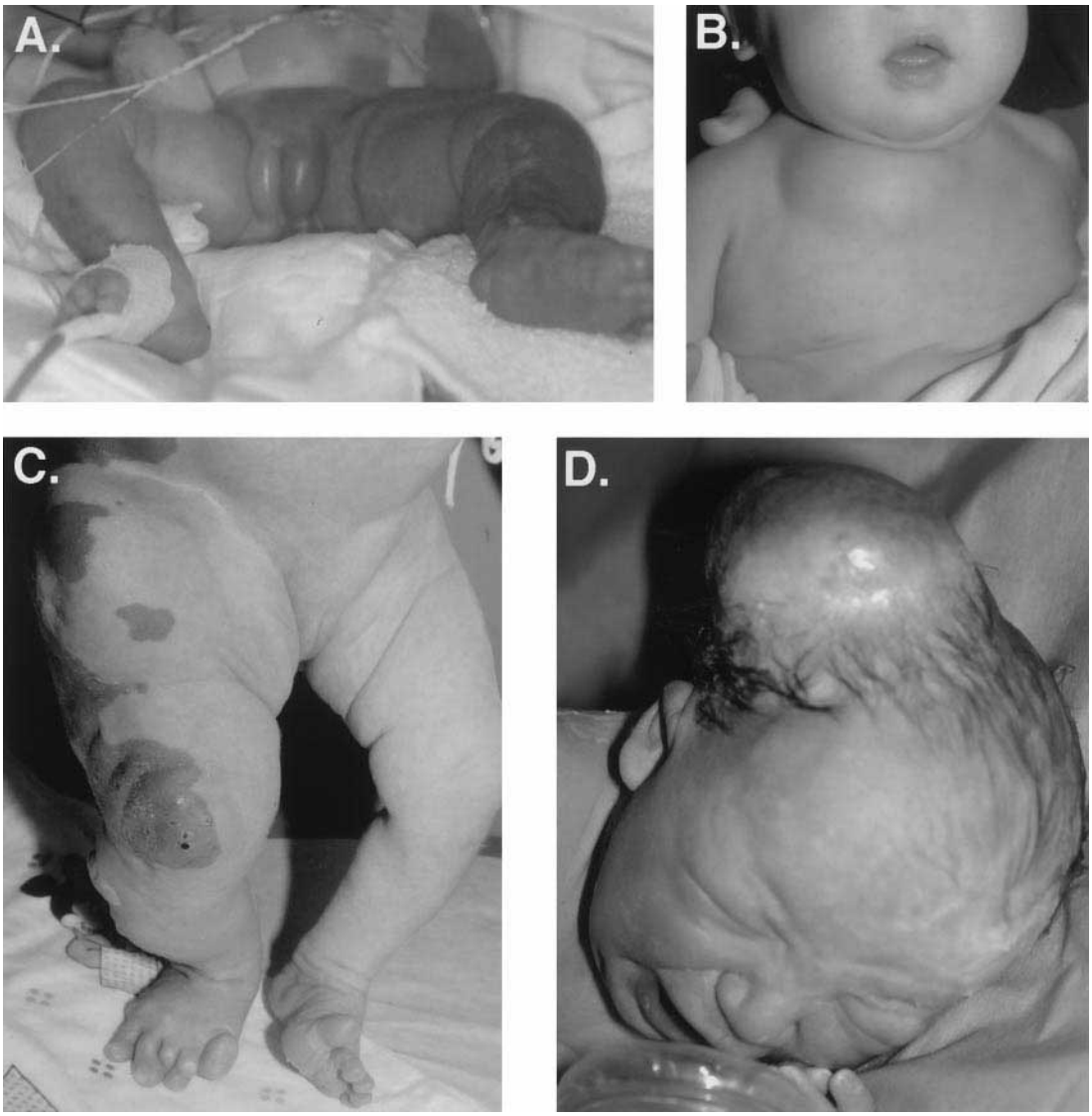

Fig 2. Examples of patients with prenatally diagnosed vascular anomalies. (A) Diffuse AVM of lower extremity and perineum of infant noted to be hydropic on antenatal sonogram (Parkes Weber syndrome). This infant died of complications of congestive heart failure. (B) Anterior cervical lymphatic malformation. (C) Capillary lymphatico-venous malformation of right lower extremity (Klippel-Trenaunay syndrome). (D) Congenital occipital hemangioma; this regressed rapidly.

resection of a hemangioma. Scalp hemangioma has a tendency to ulcerate, causing alopecia. Moreover, the scalp of an infant is lax, facilitating closure and excision, as compared with later childhood.

Hemangiomas generally pose few problems in terms of delivery or neonatal well being. We have, however, seen hepatic hemangiomas with congestive heart failure as a consequence of arteriovenous shunting or result in hypoventilation from increased intraabdominal pressure.[5] The remaining 2 correctly diagnosed hemangiomas were hepatic (1 with additional intracranial lesions). In one of these, accurate prenatal diagnosis in the fetus with evolving cardiac decompensation prompted administration of corticosteroids to the mother during the antenatal period with a successful outcome.[6] In the other, cesarean delivery was done with a neonatology team prepared to receive the infant. In both cases, accurate diagnosis contributed to optimized antenatal care.

We also have seen neonates with congestive heart failure as a result of hemangiomas with arteriovenous shunting in extrahepatic locations, although this is very rare. This is illustrated in the infant with the perineal and lower extremity hemangioma (erroneously ascribed a prenatal female gender) who had high-output cardiac decompensation during the first month of life. Multiple arteriovenous fistulae were noted on angiographic study.

The 2 other infants with incorrectly diagnosed hemangioma (the atrial hemangioma diagnosed prenatally to

be a teratoma and the hepatic neuroblastoma antenally thought to be hemangioma) underscore the importance of differential diagnosis. Accurate prenatal diagnosis in these patients might have optimized the antenatal course. Congenital fibrosarcoma also has been confused with hemangioma in the neonatal period.[7]

Vascular malformations are designated according to the predominant channel type as capillary malformation (CM), lymphatic malformation (LM), venous malformation (VM), arteriovenous malformation (AVM), and capillary-lymphatico-venous malformation (CLVM).[1] Malformations with an arterial component are rheologically fast-flow, whereas capillary, lymphatic, and venous anomalies are slow-flow in nature.

The most commonly diagnosed malformation in our series was LM, followed by CLVM. LM consists of localized or diffuse aberrantly formed lymphatic channels and cysts. The terms *cystic hygroma* and *lymphangioma* are outdated and contribute to diagnostic confusion. Sites of predilection for LM include, in order of prevalence, the cervicofacial region, axilla, mediastinum, and extremities. It is useful to designate LMs as predominantly microcystic or macrocystic on radiographic or histologic evaluation. Most LMs are obvious at birth, but they also manifest during the first 2 years of life or, occasionally, in later childhood.

Sonographic examination of an LM generally shows a cystic, anechoic mass that may be unicameral or multilocated with multiple thin septae.[8] Hemorrhagic areas within LM may be detected as hyperechoic foci within the mass. The differential diagnosis of a fetal multilocular cystic mass in the abdomen includes a congenital fibrosarcoma, cystic teratoma,[9] enteric duplication cyst, mesenteric cyst, hepatic mesenchymal hamartoma, multilocular cystic nephroma, choledochal cyst, and urachal cysts.

The differential diagnosis of a cervical cystic mass includes cystic teratoma, thymic cyst, and a congenital fibrosarcoma.[10] A clear distinction should be made between posterior nuchal translucency (PNT) seen during the first and early second trimester, and cervicofacial LM diagnosed during the second and third trimesters. The term *cystic hygroma* has caused diagnostic confusion in this regard. PNT has been referred to as *fetal cystic hygroma*, a term also applied to fetal LM.[11] PNT is associated with increased risk of aneuploidy, determined to be 62% in a review of 740 karyotyped cases.[11,12] In the presence of a normal karyotype, a fetus with a first trimester nuchal translucency has an equivocal prognosis with a normal outcome expected in 80% of pregnancies carried to the third trimester. Dysmorphic features are noted in 20% of liveborn infants, but do not include LM.[13] Normal outcome is highest when the fetal translucency resolves spontaneously, typically before 20 weeks gestation.[11] In contrast, an LM typically is noted at a later gestational age. In our series, the correct diagnosis was ascertained generally between 24 and 37 weeks (with the exception of 3 patients diagnosed at 16, 17, and 20 weeks). Cervical LM generally is anterior or lateral, and may be unilateral or bilateral, in comparison with nuchal cysts, which are invariably bilateral, symmetric, and posterior.[11]

Accurate prenatal diagnosis of an LM permits precise planning for delivery and postnatal care. The two EXIT procedures in this series, done for fetuses with large anterior cervical malformation, permitted assessment of airway patency before clamping the umbilical cord. If laryngeal LM precludes intubation, tracheostomy can be performed under controlled circumstances. In one case, intubation was possible; in the other, tracheostomy was required. Asphyxia and death have been reported in a neonate with a prenatally diagnosed cervical LM in which an EXIT procedure was not done.[14] In other cases in our series, cesarean section without an EXIT procedure was planned to avoid intrapartum complications. Lymphatic malformation has been reported as a cause of shoulder dystocia.[15]

Incorrect prenatal diagnosis of LM was associated with morbidity in this series, illustrated by unnecessary fetal surgery for the LM of the hand that was diagnosed to be “amniotic band syndrome” on prenatal sonography. It is conceivable that both LM and amniotic band syndrome were present in this patient. However, such an association has not been observed at our vascular anomalies center, nor reported in the literature.

Hydrops fetalis in association with a vascular anomaly is a poor prognostic indicator, as it is with fetal tumors.[16] In the published case reports summarized in Table 1, 3 of the 5 hydropic fetuses died; the other 2 were delivered early and survived after complicated neonatal intensive care unit courses. In our series, hydrops was seen in 1 fetus. In spite of an emergent cesarean delivery, the neonatal course was complicated by high output cardiac failure and death.

There were 2 antepartum referrals (during the reviewed interval) to our center for hydrops with fetal ascites and pleural effusion where generalized malformation of the thoracic duct or cisterna chyli was suspected. These were not included in this series because anatomic demonstration of lymphatic abnormalities was not possible. One of these infants died on day 1 of life with massive chylous ascites and chylous pleural effusions. The second infant died at the age of 1 year with recalcitrant chylous ascites and chylous pleural effusions after thoracic duct ligation, pleurodesis, and a complicated course in the intensive care unit (other anomalies also were present in this infant).

Accurate prenatal diagnosis of a vascular anomaly

permits fetal intervention, as exemplified by a fetus in this series whose cardiac decompensation was corrected after administration of maternal corticosteroids.[6] There is a reported case of a fetal LM treated with intralesional injection of OK-432.[17] It is not always clear in these cases whether fetal intervention altered the natural history of the disorder. In the case of sclerotherapy, for example, 2 posterior cysts were injected that could have been spontaneously resolving nuchal translucencies rather than a true LM. However, these examples highlight the principle of in utero therapy for vascular anomalies that threaten the life of the fetus.

This series illustrates a number of emerging concepts. Vascular anomalies can be diagnosed prenatally, typically during the second and third trimesters of pregnancy. Unfortunately, these anomalies frequently are misdiagnosed. Pediatric surgeons consulted regarding a fetal tumor should include vascular anomaly on their differential diagnostic list. Proper diagnosis facilitates optimal planning for delivery and neonatal care. Incorrect diagnosis may result in fetal morbidity and ill-advised treatment. A window for fetal intervention, including pharmacologic therapy, may open during later gestation.

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Discussion

T.M. Crombleholme (Philadelphia, PA): Very nice presentation. The fetus that you mentioned actually did have amniotic band syndrome, and the band was lysed in utero, and the lymphatic malformation that you see actually is a secondary process. We have seen this in the fetal lamb model of amniotic band syndrome. What you saw postnatally is very consistent with an amniotic band obstruction that is treated in utero.

S. Fishman (response): I appreciate that comment. I was not attempting to point fingers. We certainly considered that possibility, and I would agree that is possible. Most cases of amniotic band syndromes have atrophy distal to the band, but it certainly seems teleologically possible that constriction of an extremity could lead to lymphatic maldevelopment or hyperplasia. Histologically and radiographically, this was typical lymphatic malformation.