Detection of Intracranial Arteriovenous Fistula by Two-dimensional Ultrasonography

A. Rebecca Snider, M.D., Scott J. Soifer, M.D., and Norman H. Silverman, M.D.

SUMMARY Three newborn infants who had severe congestive heart failure caused by an arteriovenous malformation of the vein of Galen were evaluated by two-dimensional ultrasonography. Examination of the heart from standard echocardiographic planes showed right atrial and right ventricular dilatation. The superior vena cava, ascending aorta and the vessels to the head and neck were enlarged. Contrast two-dimensional echocardiography from a peripheral vein showed right-to-left atrial shunting and early superior vena cava recirculation. With the transducer positioned in the anterior fontanel or against the temporal bone, two-dimensional sector scans of the brain were obtained in the coronal, sagittal and transverse planes. A large echo-free space that represented an aneurysm of the vein of Galen was seen within the brain. During contrast injection into a peripheral vein, microcavitations that passed from right to left at the atrial level were seen filling the vein of Galen aneurysm. In two infants in whom beryllium wire was placed in the fistula to induce thrombosis, postoperative ultrasonography showed a dense mass of echoes arising from the wire and persistent left-to-right shunting through the fistula. In infants presenting in the first week of life with congestive heart failure and cyanosis of unknown etiology, two-dimensional ultrasonography provides a rapid, safe method for detecting intracranial arteriovenous malformation.

INTRACRANIAL arteriovenous malformation is a rare cause of severe congestive heart failure in infancy. Initial physical findings may include cranial bruits, hyperdynamic cardiac impulses, heart murmurs, cyanosis and bounding peripheral pulses. Because cranial bruits are often of low intensity or absent, the exact diagnosis is rarely made from the clinical examination before cardiac catheterization or death. Infants with congestive heart failure caused by intracranial arteriovenous malformations are critically ill, and prompt diagnosis is essential. When the diagnosis is suspected, cerebral angiography has been used to delineate the exact anatomy of the intracranial arteriovenous fistula. Recently, Schum and colleagues used computerized tomography to diagnose a vein of Galen fistula. To determine if two-dimensional ultrasonography could provide a rapid, noninvasive method for detecting intracranial arteriovenous malformations, we used a two-dimensional sector scanner to examine three newborn infants who had arteriovenous malformations of the vein of Galen.

Materials and Methods

Three newborn infants were studied by two-dimensional ultrasonography after a diagnosis of arteriovenous malformation of the vein of Galen was established. In cases 1 and 2, the diagnosis was made by aortography at a referring hospital; in case 3, the diagnosis was made at cardiac catheterization. In cases 1 and 2, the two-dimensional sector scan was performed before and after an attempted surgical repair, which consisted of threading beryllium wire into the aneurysm to induce thrombosis.

Two-dimensional ultrasonography was performed with a wide-angle, phased-array sector scanner (either a Varian V-3000 with a 3.5-MHz transducer or a Toshiba SSH 10A with a 2.4-MHz transducer). In each infant, a two-dimensional echocardiographic examination was performed from the standard parasternal, apical, subcostal and suprasternal notch planes. In addition, two-dimensional ultrasound examination of the brain was performed in each infant. To examine the brain in multiple coronal or sagittal planes, the transducer was positioned in the anterior fontanel and swept anteroposteriorly in a coronal body plane and from right to left in a sagittal body plane. A transverse view of the brain was obtained by applying the transducer to the temporal bone in a horizontal plane. Two-dimensional contrast ultrasonography was obtained in all cases by the injection of 1–2 ml of sterile saline into an arm vein.

The two-dimensional sector scans were recorded on videotape for later analysis in real-time or slow motion. The figures presented here were taken from Polaroid photographs of stop-action, single-frame images from the videotape recordings, and lack the integrated image and visual appreciation of motion normally present in the real-time recordings.

Case Reports

Case 1

A 2780-g, full-term female infant was born after normal pregnancy, labor and delivery. Symptoms of congestive heart failure appeared soon after birth, and
a cardiac consultation was obtained. Physical examination showed a mildly cyanotic infant in moderate respiratory distress. The peripheral pulses were bounding in all extremities, and there was no radial-femoral delay. A right ventricular heave was present at the lower left sternal border. The first heart sound was normal; the second heart sound was narrowly split with an increased pulmonic component. A grade 2/6 long systolic murmur was heard at the lower left sternal border and radiated to the lower right sternal border. A grade 2/4 middiastolic murmur was heard at the lower left sternal border. A gallop rhythm was present. A grade 1–2/6 continuous murmur was heard over the cranium, especially posteriorly.

The ECG showed biventricular hypertrophy. The chest x-ray showed generalized cardiomegaly with increased pulmonary vascular markings. M-mode echocardiography showed right ventricular dilatation and a dense pulmonic valve echo with midsystolic closure. A carotid arteriogram performed because of the cranial bruit showed a large arteriovenous malformation of the vein of Galen. The infant was transferred to the University of California, San Francisco, for surgery. An occipitoparietal craniotomy was performed when the patient was 3 days old, and 100 feet of beryllium wire were introduced into the vein of Galen aneurysm to induce thrombosis. Postoperatively, the infant’s course was complicated by persistent congestive heart failure, renal failure and neurologic impairment. The infant died at age 44 days.

Case 2

A 3500-g, full-term female infant was delivered by cesarean section after a normal pregnancy. Cyanosis and respiratory distress developed soon after birth. On physical examination, a cranial bruit was audible over the anterior fontanel and right parietal region. The peripheral pulses were bounding in all extremities. A right ventricular heave was felt at the lower left sternal border. The first heart sound was normal; the second heart sound was narrowly split, with an accentuated pulmonic component. A grade 2/6 long systolic murmur was heard at the lower left sternal border and a gallop rhythm was present.

The ECG showed biventricular hypertrophy. The chest x-ray showed generalized cardiomegaly with increased pulmonary vascularity. M-mode echocardiography showed evidence of right ventricular dilatation. There was midsystolic closure of the pulmonic valve. Cerebral angiography performed because of a cranial bruit showed a large arteriovenous malformation of the vein of Galen. The infant was referred to the University of California, San Francisco, for surgery. At surgery, beryllium wire was introduced through a craniotomy into the arteriovenous malformation to induce thrombosis. Postoperatively, the infant showed persistent congestive heart failure unresponsive to medical management. Three other surgical attempts were made, all unsuccessfully, to clip arterial feeding vessels communicating with the fistula and introduce more beryllium wire; the infant died at age 4 months.

Case 3

A 3200-g, full-term male infant was delivered after an uncomplicated pregnancy. Apgar scores were 2 and 6 at 1 and 5 minutes. By 12 hours of age, the infant developed severe congestive heart failure and was transferred to the University of California, San Francisco, for cardiac evaluation. Physical examination showed a cyanotic male infant in marked respiratory distress. The peripheral pulses were diminished in all extremities. A right ventricular heave was present at the lower left sternal border. The first heart sound was normal; the second heart sound was single and accentuated. No cardiac murmurs were heard, and a gallop rhythm was present. There was marked hepatomegaly.

The ECG was normal. The chest x-ray showed severe cardiomegaly and increased pulmonary vascular markings. The M-mode echocardiogram showed a large right ventricle and a thick pulmonary valve echo with midsystolic closure. At 36 hours of age, the infant underwent right- and left-heart catheterization, which showed a large arteriovenous malformation of the vein of Galen, pulmonary artery hypertension, a patent ductus arteriosus and a patent foramen ovale. A computerized tomographic scan of the brain showed the arteriovenous malformation and enlarged ventricles. After catheterization, a systolic murmur was heard on both sides of the cranium. The infant’s course continued to deteriorate despite medical management, and he died at age 4 days.

Results

Cardiac Scans

Two-dimensional echocardiograms in all three infants showed right atrial and right ventricular dilata-

![Figure 1. Two-dimensional echocardiogram in the suprasternal notch short-axis plane from patient 1. There is marked enlargement of the superior vena cava (SVC) caused by increased systemic venous return from the vein of Galen aneurysm. Ao = aorta; LA = left atrium; PA = pulmonary artery; R = right; S = superior; I = inferior; L = left.](image-url)
tion. The main pulmonary artery was enlarged and the pulmonary valve was densely reflective in all three patients. The left atrium, left ventricle, aortic valve and septal motion were normal. A long-axis view of the aortic arch from the suprasternal notch in all three infants showed that the ascending aorta and carotid arteries were dilated. In addition, a short-axis view of the aortic arch from the suprasternal notch showed marked enlargement of the superior vena cava and innominate veins (fig. 1).

Contrast two-dimensional echocardiograms in the apical four-chamber projection showed microcavitations from the contrast injection passing from the right atrium to the left atrium via a patent foramen ovale. In addition, contrast two-dimensional echocardiography was performed with the transducer positioned in the suprasternal notch in a coronal body plane (fig. 2). After the contrast injection, microcavitations were seen first in the superior vena cava and then, nearly simultaneously, in the right pulmonary artery and left atrium because of forward flow in the right heart and atrial right-to-left shunting. A short time later, contrast reappeared in the superior vena cava because microcavitations that reached the ascending aorta via a right-to-left intracardiac shunt were able to pass from left to right within the intracranial arteriovenous malformation and recirculate to the superior vena cava.

Patients 1 and 2 were reevaluated postoperatively by contrast two-dimensional echocardiography. Both patients showed continued right-heart dilatation, thick pulmonary valve echoes and right-to-left atrial shunting. These findings were present in patient 2, 5 weeks postoperatively.

**Brain Scans**

In the coronal, sagittal and transverse planes, a large fluid-filled structure was imaged within the brain (figs. 3–5). In patient 3, the structure extended more anteriorly than in patients 1 and 2. A contrast injection of 1–2 ml of sterile saline was made in an arm vein while the coronal, sagittal or transverse plane of the head was imaged. Microcavitations that passed from right to left at the atrial level traveled in the as-

![Contrast two-dimensional echocardiograms in the suprasternal notch short-axis plane from patient 2. (1) Before contrast injection. The superior vena cava (SVC) is large. AO = aorta; LA = left atrium; PA = main pulmonary artery; R = right; L = left; S = superior; I = inferior. (2) Microcavitations (arrows) from a left arm vein injection are seen in the SVC. (3) Microcavitations have completely opacified the SVC and PA and have passed into the LA and AO (arrows) via a right-to-left atrial shunt. (4) The AO, PA, and LA are relatively free of echoes from the contrast injection; however, microcavitations (arrows) have reappeared in the SVC. The SVC recirculation is caused by microcavitations passing from left to right through the vein of Galen aneurysm.](http://circ.ahajournals.org/content/70/6/1181.full.html)
Figure 3. (top) Two-dimensional sector scan of the brain in the coronal plane in patient 3. The lateral ventricles (V) are seen as echo-free spaces. The echo-free space posterior to the lateral ventricles is the vein of Galen fistula (F). I = inferior; S = superior; L = left; R = right. (middle) Contrast two-dimensional sector scan in the coronal plane in patient 3. Microcavitations from the contrast injection (white arrows) are seen in the fistula. (bottom) Computerized tomographic scan in the coronal plane in patient 3. The dense white structure in the center of the brain is the vein of Galen fistula opacified by a contrast agent. The lateral ventricles are enlarged.

Figure 4. (top) Two-dimensional sector scan of the brain in the sagittal plane in patient 3. The lateral ventricle (V) is seen as an echo-free space. The echo-free space inferior to V is the vein of Galen fistula (F). A = anterior; P = posterior; I = inferior; S = superior. (middle) Contrast two-dimensional sector scan in the sagittal plane in patient 3. Microcavitations from the contrast injection (white arrows) are seen in the fistula. (bottom) Computerized tomographic scan in the sagittal plane of patient 3. The vein of Galen fistula is opacified by a contrast agent.
which were nonvascular and therefore not opacified by contrast echoes (figs. 3–5).

The exact anatomy of the afferent and efferent vessels connecting to the arteriovenous fistula could not be defined by two-dimensional ultrasonography. However, a large anterior cerebral artery connecting to the vein of Galen aneurysm was imaged in patient 1. During contrast injections, microcavitations passed through this anterior cerebral artery into the vein of Galen fistula (fig. 6).

Two-dimensional sector scans of the brain were obtained postoperatively in patients 1 and 2 (fig. 7). In the coronal, sagittal and transverse planes, a mass of dense echoes was seen in the area previously occupied by the fluid-filled fistula. The dense echoes arose from the mass of beryllium wire threaded into the fistula at the time of surgery. In both patients, echo-free areas were seen within these dense echoes. These echo-free areas represented residual blood-filled areas in the vein of Galen fistula. During postoperative contrast injections in patients 1 and 2, microcavitations were seen in these echo-free areas because of continued left-to-right flow in the fistula.

Discussion

Besides the complex adjustments that occur on conversion to extraterine circulation,8 the newborn infant with a large intracranial arteriovenous malformation has an additional circulatory burden. Shortly after birth, when the pulmonary vascular resistances are still elevated, the decrease in the total systemic vascular resistance caused by the presence of a large arteriovenous malformation promotes right-to-left ductal shunting. The large venous return to the right atrium from the arteriovenous fistula augments right-to-left atrial shunting. In the presence of a large right-to-left ductal shunt, the pulmonary blood flow and left atrial volume are decreased, and further right-to-left atrial shunting occurs because the flap valve of the foramen ovale remains open.9

This persistence of fetal circulatory patterns allows microcavitations from a two-dimensional contrast injection to bypass filtration in the pulmonary capillary bed and gain access to the systemic circulation. The passage of microcavitations from the systemic circulation into the low-resistance fistula can be imaged by two-dimensional ultrasonography of the brain. Microcavitations in the fistula bypass filtration by the systemic capillary bed and reappear rapidly in the
FIGURE 6. (top) Two-dimensional sector scan of the brain in the sagittal plane in patient 1. The large echo-free space represents the vein of Galen fistula (F). An anterior cerebral artery (CA) communicates with the fistula. A = anterior; P = posterior; I = inferior; S = superior. (middle) Microcavitations from a peripheral contrast injection are seen entering the fistula from the area of the CA. (bottom) Most of the fistula is opacified by echoes from the contrast injection.
to dilatation of the ascending aorta and carotid arteries, which can also be detected by two-dimensional suprasternal notch echocardiography. Increased systemic venous return from the arteriovenous fistula leads to superior vena caval, right atrial and right ventricular dilatation, also detectable by two-dimensional echocardiography.

An intracranial arteriovenous fistula in a newborn infant with cyanosis and congestive heart failure may be difficult to recognize clinically. In 1972, Holden and colleagues reported that the diagnosis was made in only four of 26 such infants before death. In 1976, Watson and colleagues reported that in less than half of infants with congestive heart failure due to vein of Galen fistula was the diagnosis made during life. The clinical diagnosis is usually made if a cranial bruit is detected; however, cranial bruits may be of low intensity, absent or missed during the initial physical examination. Because infants with congestive heart failure and cyanosis caused by an arteriovenous malformation are usually critically ill, rapid diagnosis is essential. We found that two-dimensional ultrasonography allows accurate, rapid detection of the intracranial arteriovenous malformation but does not provide the detailed anatomic delineation of the feeding vessels that is necessary before surgery. If the diagnosis can be made by ultrasonography without cardiac catheterization, both the additional stress of catheterization and use of contrast agents at catheterization can be avoided. Once the diagnosis is made by two-dimensional ultrasonography, the use of contrast agents can be reserved for more detailed anatomic studies, such as selective cerebral arteriograms in several views or computerized tomography with contrast in several planes before surgery.

In infants presenting in the first week of life with congestive heart failure and cyanosis of unknown etiology, two-dimensional ultrasonography of the heart and brain with saline contrast injections provides a rapid, efficient method for the detection of intracranial arteriovenous malformations. Two-dimensional contrast ultrasonography also provides a safe method for the postoperative detection of persistent intracardiac right-to-left shunting and intracerebral left-to-right shunting in these infants.

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References

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A R Snider, S J Soifer and N H Silverman

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