Congestive Heart Failure and Absent Femoral Pulses in Newborns without Coarctation of the Aorta

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Two infants with thrombosis of the abdominal aorta are discussed. In each case the presentation was indistinguishable from that in coarctation of the aorta, with heart failure and absent femoral pulses. Surgery in one infant successfully relieved the obstruction. The diagnosis may not be suspected from the history. Aggressive management is indicated.

Vascular occlusion in the newborn is an uncommon clinicopathologic entity associated with significant morbidity and mortality. Several reports have described the development of aortic thrombosis in infants with a history of umbilical artery catheterization and have demonstrated the high mortality associated with this complication. We present two additional cases of aortic occlusion in infancy, emphasizing the clinical presentation as being indistinguishable from that of severe coarctation of the aorta, and demonstrating that aggressive therapy may be successful.

Case Reports

Case 1

An eight-day-old male had been born by cesarean section at term to a mother with gestational diabetes mellitus. Birth weight was 3,300 g, and Apgar scores were 4 and 8 at one and five minutes, respectively. He was meconium stained, but was not intubated, and the umbilical vessels were not catheterized. He was discharged at five days of age. Two days later respiratory distress developed, and he was transferred to Children's Hospital of Pittsburgh. The systolic pressure was 56 mm Hg in the right arm and 30 mm Hg in the left leg. There was no murmur, the liver was enlarged, and the femoral pulses were absent. Chest roentgenogram showed cardiomegaly and increased vascular markings. The ECG was normal. M-mode echocardiogram showed diminished left ventricular shortening fraction. Hemoglobin was 16.6 g/dl and hematocrit 51.5 percent; the WBC count was 12,700/cu mm, with polymorphonuclear preponderance; the platelets were 120,000/cu mm; PT was 12 seconds (control, 11.5 seconds), and PTT 36 seconds (control, 34 seconds). The clinical impression was coarctation of the aorta and congestive heart failure. Mechanical ventilation, digoxin, and prostaglandin E, infusion were begun, but the infant showed little improvement, and cardiac catheterization with angiography was performed. The study showed a normal aortic arch, a patent ductus arteriosus, and no coarctation. Flow in the descending aorta was sluggish, and the distal aorta was not visualized. The impression was thrombotic occlusion of the abdominal aorta. Despite heparinization and intensive medical efforts, the infant died 19 hours after admission.

At autopsy a large thrombus was found occluding the abdominal aorta from a point just proximal to the renal arteries to the bifurcation of the aorta (Fig 1). The thrombus propagated into the left renal artery, and small recent infarcts were present in the left kidney. No inflammation or degeneration was noted in the adjacent aortic wall, and no venous thrombi were present.

Case 2

A 3,700-g male was born at term to a mother with gestational diabetes mellitus. Vaginal delivery was complicated by shoulder dystocia, with asphyxia and Apgar scores of 1 and 1 at one and five minutes, respectively. Umbilical artery and vein catheters were inserted, bicarbonate was administered (exact route not known), and the infant was transferred to another hospital. The hemoglobin prior
to transfer was 12.9 g/dl. In the first day of life, the left leg became cyanotic, but this cleared after immediate removal of the catheter. Seizures were controlled with phenobarbital and phenytoin, and the infant's status improved until the fifth day of life, when both legs became transiently cyanotic and the femoral pulses diminished. During the subsequent four days, the femoral pulses became more difficult to appreciate, and on the tenth day of life, signs of congestive heart failure and pulmonary hemorrhage developed. Digoxin and antibiotic therapy, and mechanical ventilation were begun, and the infant was transferred to St. Christopher's Hospital for Children. On examination, the infant was hypertonic with occasional spontaneous respirations and frequent seizure activity. Systolic pressure was 140 mm Hg in the left arm and 50 mm Hg in both legs. Small amounts of blood were suctioned from the endotracheal tube. A continuous murmur was present at the left infraclavicular space, the liver was enlarged, and the femoral pulses were absent. The ECG showed biventricular hypertrophy, and a roentgenogram of the chest showed cardiomegaly, with parenchymal haziness and scattered densities compatible with congestive heart failure and pulmonary hemorrhage. A two-dimensional echocardiogram failed to satisfactorily visualize the distal aortic arch. Hemoglobin was 13.6 g/dl and hematocrit, 40.8 percent. The white blood cell count was 14,000/mm^3, with polymorphonuclear preponderance; and platelets, 58,000/cu mm. There was occult hematuria. An infusion of prostaglandin E, was begun, but six hours later the infant's status was not improved, and cardiac catheterization with angiography was performed. The study showed a normal aortic arch and complete occlusion of the distal abdominal aorta proximal to the ostia of the renal arteries (Fig 2). Thrombolytic therapy was not initiated because of signs of a bleeding diathesis, but in view of hypertension and CNS irritability, surgical embolectomy was attempted via a transverse lower abdominal incision and transperitoneal approach. The abdominal aorta was opened just above the level of the bifurcation, and a large thrombus was removed. Smaller thrombi were also removed from both iliac arteries with Fogarty catheters. There was prompt resolution of the systemic hypertension, and appearance of excellent pulses in the lower extremities. Heparin therapy was begun eight hours after surgery and was continued for three days. Neurologic problems persisted which required adjustment of medications. Seven weeks after surgery, the infant was discharged with normal blood pressure and pulses.

**DISCUSSION**

Thrombotic occlusion of the abdominal aorta in infants is rare, and has been observed in association with umbilical artery catheterization, as well as in cases where the umbilical vessels had not been manipulated. Sepsis, polycythemia, maternal diabetes, dehydration, and clotting factor abnormalities have been considered to play a role in the etiology. In those patients with no history of umbilical artery catheterization, it has been suggested that thromboembolic phenomena may be related to the changes in the circulation at birth, with emboli originating in a patent ductus arteriosus or in the umbilical vein.

Our two patients illustrate the typical presentation and rapid deterioration characteristic of this disorder. The nature of the problem in case 1 was unsuspected, while in case 2 it was suspected from the history. In both cases the role of maternal diabetes and perinatal asphyxia in the development of aortic thrombosis must remain speculative, but in case 2 the umbilical artery catheter and hypertonic fluid infusions must be considered contributing factors. Polycythemia was not present in either infant. Although in our cases the diagnosis was confirmed by angiography, other investigators have used Doppler blood flow and echocardiography as well as radionuclide perfusion studies to diagnose the condition noninvasively, thus avoiding the use of angiographic contrast media that could decrease the chances of survival.

Although conservative management has proven successful in cases of peripheral occlusive disorders in infants, we are not aware of survival of any infant with symptomatic occlusion of the abdominal aorta with medical therapy alone. It is possible that thrombolytic therapy may provide an alternative to the management of these critically ill infants, but success may depend on early recognition and diagnosis. It is questionable whether thrombolytic therapy could have been successfully administered in case 2 in view of signs of pulmonary hemorrhage. The value of postoperative anticoagulation or thrombolytic therapy remains to be defined.

The clinical course of these two infants demonstrates that thromboembolic aortic occlusion must be suspected in any infant who presents with signs suggestive of coarctation of the aorta, and that aggressive surgical management of the symptomatic infant can successfully relieve the obstruction.

**ADDENDUM**

Since submitting this manuscript, a report has appeared describing the failure of intra-arterial thrombolytic therapy to induce clot lysis in a newborn with aortic occlusion. Failure was attributed to inability to induce a hyperfibrinolytic state due to physiologic reduction in plasminogen levels.
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REFERENCES


Paroxysmal Nodal Reentrant Tachycardia*

Surgical Cure with Preservation of Atrioventricular Conduction


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In a patient with repetitive disabling tachycardias refractory to pharmacologic treatment, the electrophysiologic study suggested the existence of atrioventricular nodal reciprocating tachycardia. During ventricular pacing, endocardial mapping of the lower atrium showed the atrial breakthrough point in an area of the lower interatrial septum close to the AV node crista. A selective atriotomy was performed. The postoperative electrophysiologic studies showed absence of ventriculoatrial conduction at several ventricular pacing rates, while antegrade conduction is preserved. The patient remained free of arrhythmias 21 months after surgery, taking no antiarrhythmic drugs.

Electrophysiologic studies using programmed stimulation,1-3 endocardial mapping,4-5 and surgical results6,7 have led to a better knowledge of AV junctional reciprocating tachycardias. There is more experience in the surgical treatment of AV junctional reciprocating tachycardias incorporating Kent bundles in the circuit. However, those tachycardias with intranodal mechanism or those incorporating other types of accessory pathways are less well known, and surgical treatment experience is limited.8,11

The surgery in the latter type of AV reciprocating tachycardias basically depends on the surgical interruption or cryoablation of the His bundle, as demonstrated by Harrison et al9 and Klein et al.10 Pritchett et al10 and Ward et al11 have recently reported surgical correction of arrhythmia while preserving AV conduction.

We recently operated on a patient with recurrent AV reciprocating tachycardia which was refractory to medical treatment, using endocardial mapping of the lower atrium close to the AV junction prior to AV interruption of the tachycardia circuit. The patient remained free of arrhythmias 21 months after surgery, with preserved AV conduction.

CASE REPORT

The patient was a 62-year-old woman with no clinical or hemodynamic evidence of heart disease, who had had bouts of palpitations since 1966. Initially, the tachycardia appeared once or twice a month, lasted for 15 to 30 minutes, and remitted spontaneously. Since 1975 the tachycardia episodes had become more frequent (five to six per month) and lasted longer. From 1979 on, the crises increased to three or four per week, ranging from three to 12 hours, with occasional syncope. Furthermore, the patient showed evidence of severe impairment of her quality of life and was unable to perform her usual activities.

The arrhythmia remitted temporarily after intravenous (IV) administration of verapamil (10 mg), ajmaline (1 mg/kg), or mexiletine (250 mg as single doses). Multiple oral antiarrhythmic drugs were used without preventing tachycardia, the only ones showing some effect being amiodarone (600 mg/day) and quinidine sulfate (1 g/day), or the association of digoxin (0.25 mg/day) with verapamil (240 mg/day), which lengthened the intercrisis interval and shortened the crises.

Preoperative Electrophysiologic Study

This study was carried out to assess the mechanism of arrhythmia and the action antiarrhythmic drugs and diverse modalities of cardiac pacing.

The basal ECG showed sinus rhythm with 0.12-sec P-R interval, left anterior hemiblock and occasionally right bundle branch block (Fig 1A). All antiarrhythmic medication was suspended one week before the study. Three multipolar electrode catheters were inserted.