RADIOLoGY

Single Film Retrograde Umbilical Aortography in the Diagnosis of Hypoplastic Left Heart Syndrome with Aortic Atresia

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SUMMARY Single film retrograde umbilical aortography was used successfully in confirming the diagnosis of hypoplastic left heart in three neonates. The method involves injecting Renografin 75% (1.25 cc/kg body weight) into the umbilical artery catheter according to the following sequence. First, the tip of the catheter is positioned at the level of the ductus arteriosus. The infant heart beats are then counted from one to three. The contrast media injection is begun manually on the count of one and an anterior-posterior chest film is taken on the count of three. Thus the film is taken one diastolic period after completion of the injection.

The proper timing of the contrast media injection and filming is essential for obtaining a diagnostic film. This procedure was diagnostic in the three infants studied and eliminated the need of transferring two sick infants from affiliated hospitals.

NEONATES with hypoplastic left heart syndrome, despite a number of attempts at different palliative procedures, continue to have a dismal outlook with almost all such babies succumbing in the first week of life. When these critically ill infants are first seen, it is important to exclude a more remediable lesion. The definitive diagnosis is frequently made with cardiac catheterization and angiography, although cardiac catheterization laboratory facilities for sick infants are not available in all hospitals thus necessitating transfer of these babies. Echocardiography is another useful diagnostic tool, although it, too, is unavailable in some hospitals.

In this report we describe a simple and rapid method by which the diagnosis of hypoplastic left heart syndrome was established in three newborn infants in the intensive care unit. This was accomplished by single film retrograde aortography injection via an umbilical artery catheter.

Case Report

A full term 2920 gram male infant was transferred to the UCLA Medical Center at 36 hours of age because of tachypnea and cyanosis. He was the product of an uneventful 38-week gestation and normal delivery. Symptoms were first noted at 12 hours of age. Due to the presence of severe metabolic acidosis he was given a total of 8 cc sodium bicarbonate prior to the transfer. Physical examination revealed a critically ill, pale infant in marked respiratory distress. His heart rate was 160/minute; his respiratory rate was 72/minute. The precordium was hyperactive and the peripheral pulses were unobtainable. The first and second heart sounds were loud, and there were no murmurs. The liver was 6 cm palpable below the right costal margin. The electrocardiogram showed severe right ventricular hypertrophy with depressed ST segment in the right precordial leads and decreased left ventricular forces. The chest roentgenogram revealed moderate cardiomegaly associated with pulmonary venous congestion. The clinical diagnosis of probable hypoplastic left heart syndrome was made. Because of the late hour and before mobilizing the emergency cardiac catheterization team, it was decided to obtain a single film aortogram in the neonatal intensive care unit in an attempt to confirm the clinical diagnosis and possibly obviate the need for cardiac catheterization.

A #5 Fr Argyle catheter was inserted into the umbilical artery and advanced retrograde into the aorta. The tip of the catheter was positioned above the diaphragm between the fourth and sixth thoracic vertebrae and confirmed by a chest roentgenogram (18-20 cm from the umbilicus in a full term infant).

Anterior-posterior chest film was taken after injecting Renografin 75% (1.25 cc/kg body weight) into the aortic catheter according to the following sequence: The infant cardiac beats were counted on the scope from one to three. Three beats on the average takes from one to 1.2 seconds. The infant was positioned for anterior-posterior chest film. A rapid manual injection was begun on count one and the film was taken on count three. Thus the film was taken two diastolic periods after beginning the injection and one diastolic period after completion of the injection.

The aortogram (fig. 1) demonstrated retrograde filling of the aortic arch and neck vessels which appeared normal. There was also opacification of a hypoplastic ascending aorta and faint opacification of the right coronary artery. At the level of the aortic isthmus a ridge was seen as might be found in coarctation of the aorta. The procedure took 4-5 minutes after positioning the aortic catheter.

This procedure was performed by one of the authors at an affiliated hospital in two additional neonates with hypoplastic left heart syndrome. The angiographic findings were similar to the case described above. All three infants tolerated the injection well. Postmortem examination in all three infants confirmed the diagnosis of hypoplastic left heart syndrome with mitral and aortic atresia.
Discussion

Many neonates with serious congenital heart disease die within the first four weeks of life. In recent years, more aggressive medical and surgical management resulted in the survival of many of these infants. One invariably lethal condition is the hypoplastic left heart syndrome with aortic atresia. Although the clinical and laboratory findings described in our first case were highly suggestive of the hypoplastic left heart syndrome, the definitive diagnosis should be made as soon as possible to identify other more remediable lesions such as complicated forms of coarctation of the aorta with heart failure; critical aortic stenosis; total anomalous pulmonary venous return; forms of transposition of the great arteries; and occasionally so-called "persistence of the fetal circulation" with poor left ventricular function.

Echocardiography is a safe, noninvasive technique, which is used effectively in the diagnosis of many of these defects, and should be utilized first in evaluating infants with congenital heart disease. Although the diagnosis of hypoplastic left heart is made in many infants with echocardiography, angiography is used frequently to confirm or delineate the diagnosis when the echo findings are questionable or inconclusive. The expertise and equipment to perform echocardiograms in sick infants are indeed available to the large pediatric cardiology centers, but are not available to small pediatric cardiology centers, or to newborn nursery units with only one pediatric cardiologist on their staff. Furthermore, cardiac catheterization carries high risk in these sick infants because of poor cardiac and pulmonary function, transferring the baby to the cardiac catheterization laboratory or to the cardiac center, significant drop in temperature during transfer, and blood loss during cardiac catheterization. Furthermore, aortogram is the most important diagnostic procedure during cardiac catheterization in these infants. For these reasons it is desirable to have another simple procedure for confirming the expected diagnosis of hypoplastic left heart syndrome with aortic atresia. A single film aortogram was successful in delineating the diagnosis in the three infants studied, and eliminated the need for transferring the two sick infants to a larger center. Thus it appears that this technique is useful in establishing the diagnosis of aortic atresia with hypoplastic left heart syndrome.

The procedure should be performed by a pediatric cardiologist and/or pediatric radiologist with experience in angiography. It is both safe and diagnostic when done by experienced persons, and precludes the need for mobilizing a large cardiac catheterization team which is needed for safe cardiac catheterization in infants. Proper timing of injection and filming is essential for a successful single film aortogram. When the injection was made quickly and begun two beats before filming we were successful in opacifying well the ascending aorta.

It might be argued that in using this method, the diagnosis of a normal sized left ventricle, normal mitral valve and ventricular septal defect will be missed in the presence of aortic valve atresia and hypoplastic ascending aorta, and that palliative surgery is possible in this condition. However, a normal sized left ventricle with aortic atresia is a rare condition and is accompanied by normal or increased left ventricular forces on the electrocardiogram, and different findings on physical examination from those of hypoplastic left heart. In such a case, we recommend that cardiac catheterization and not a simple aortogram be performed. In the classical hypoplastic left heart with aortic and mitral atresia, palliative cardiac surgery has been unsuccessful although surgical attempts should be continued in some large medical centers. These procedures are not yet practical in all medical centers.

This method was also used successfully in our neonatal intensive care units to evaluate a large number of preterm infants with respiratory distress syndrome complicated by a patent ductus arteriosus.

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References

5. Lundstrom NR: Ultrasound cardiographic studies of the mitral valve region in young infants with mitral atresia, mitral stenosis, hypoplasia of the left ventricle and cor triatriatum. Circulation 45: 324, 1972
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