Prenatal diagnosis and postnatal management of critical aortic stenosis

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ABSTRACT Fetal echocardiography has yet to have an impact on the treatment of congenital heart disease. Critical aortic valve stenosis was diagnosed by echocardiography before birth in a 35 week gestation fetus. The risks to the fetus and mother associated with prolonged rupture of membranes prompted their transport to a hospital with cardiac surgical and high-risk perinatal facilities. Prenatal and postnatal echocardiographic findings agreed regarding the diagnostic criteria for critical aortic stenosis and primary forms of cardiomyopathy were excluded. Results of fetal Doppler examination were consistent with valvular aortic stenosis and excluded mitral regurgitation. Determination of the left ventricular size excluded ventricular hypoplasia. The infant was delivered by cesarean section and underwent successful emergency aortic valvotomy at 12 hr of age. Fetal echocardiography, in combination with a multidisciplinary postnatal approach, can be used in the successful treatment of a severe form of congenital heart disease. Circulation 75, No. 3, 573–576, 1987.

TECHNIQUES of fetal echocardiography are now being evaluated to define their accuracy in prenatal diagnosis of congenital heart disease. Early results show that defects that are discovered in utero often are severe and are associated with either other major defects, such as hydrocephaly, omphalocele, or a genetic abnormality, or with insufficiency of the atrioventricular valve, hydrops fetalis, or complete heart block. The natural history of congenital heart defects diagnosed by fetal echocardiography has yet to be fully explored, but the high mortality in this selected group has led to pessimism about the possible benefits of efforts in this field.1,2 Therefore, the major impact of fetal echocardiography up to the present time has been (1) reassurance for families to which a previous child with a complex defect has been born when the fetal echocardiographic findings appear normal, and (2) screening for congenital heart disease if a defect is suspected early in gestation. This report shows how fetal echocardiography may affect the treatment of congenital heart disease.

Methods

Prenatal studies. A gravida II, para I, Latin American mother who had received little prenatal care presented to Jefferson Davis Hospital with leaking membranes at 35 weeks estimated gestation. Initial ultrasonography showed markedly reduced amounts of amniotic fluid and a viable fetus with an estimated weight of 1900 g. The heart was noted to be abnormal and “cardiomyopathy” was suspected. There were no signs of labor. Two days later, repeat ultrasonography indicated the presence of endocardial fibroelastosis. There were no signs of fetal distress and the biophysical profile was normal. Steroids were administered to induce fetal lung maturity.

On the fourth day after admission an evaluation by a pediatric cardiologist was obtained because of the likelihood that delivery would be necessary since the mother had a low-grade fever and an elevated white blood cell count. An echocardiographic examination was done.

Echocardiographic techniques. The fetal echocardiographic evaluation was performed with the use of the ATL Ultramark 8 and the Acuson 128 dynamically focused ultrasound scanner. Pulsed Doppler fetal examination was performed by previously described techniques.3 Doppler ultrasonic intensities were limited to below 100 mW/cm² spatial peak time average. Prenatal diagnostic studies were compared with echocardiographic and operative findings postnatally.

Postnatal surgery. Respiratory distress developed in the neonate at 6 hr of age and required assisted ventilation and prostaglandin E₁ infusion. Open aortic valvotomy was performed at 12 hr of age with the use of normothermic cardioul-
Adult mortality and the presence of a severely stenotic aortic valve was confirmed. The postoperative course was complicated by the development of clinical hyaline membrane disease at 24 hr of age, which resolved over the next 48 hr. The postoperative echocardiogram showed good left ventricular function with severe hypertrophy and a closed ductus arteriosus (prostaglandin E1 had been discontinued after aortic valvotomy). There was evidence of residual aortic stenosis without insufficiency.

**Results**

Fetal echocardiography showed the fetal heart rate to be 130 beats/min and there was no arrhythmia. The left ventricle appeared to be thickened, with a cross-sectional area in the long axis projection of 1.8 cm² (normal-term newborn values compatible with perinatal survival ≥1.7 cm²). There was relative enlargement of the right ventricle and pulmonic valve and mild hypoplasia of the aortic valve anulus, which measured 5 mm in diameter (figure 1). The aortic valve appeared abnormal and stenotic. The mitral and tricuspid valves appeared intrinsically normal, although the mitral valve had reduced excursion. Shortening of the left ventricle was undetectable. There was increased echogenicity of the papillary muscles of the mitral valve. The ascending aorta was dilated and measured 9 mm in diameter at its widest point. Sampling of the ascending aorta was difficult and a maximum systolic velocity of 1.7 m/sec (normal 1.2) was obtained. There was no mitral regurgitation. The findings, by criteria for diagnosis in infancy, were consistent with critical aortic valve stenosis.

After evaluation of the relative fetal and maternal risks of a vaginal delivery, the mother was transferred to St. Luke’s Episcopal Hospital, where an elective cesarean section was performed. The infant weighed 2070 g and had Apgar scores of 8 and 9 at 1 and 5 min, respectively. There was no respiratory distress. After admission to the Texas Children’s Hospital Neonatal Intensive Care Unit, an echocardiogram confirmed the diagnosis of critical aortic valve stenosis based on the presence of (1) an abnormal aortic valve with doming, (2) abnormal left ventricular wall thickness with reduced shortening fraction, (3) right ventricular and main pulmonary arterial enlargement, (4) increased echogenicity of the mitral papillary muscles, (5) poststenotic dilation of the ascending aorta with an aorto-valve ratio of 1.8, and (6) a continuous-wave Doppler–measured velocity across the aortic valve of 3.6

**FIGURE 1.** Ultrasonography of a fetus with critical aortic valve stenosis at 35 weeks gestation. Imaging of the aortic valve and the ascending aorta (top) show the relative poststenotic dilation of the ascending aorta (5 mm diameter aortic valve anulus and 9 mm diameter ascending aorta). The four-chamber view shows the relative thickening of the left ventricular chamber, enlargement of the right ventricular chamber, and echodensity of the mitral papillary muscle (open white arrow in bottom). AAo = ascending aorta; LV = left ventricle; RV = right ventricle.
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proving the preoperative condition in infants with this disease have been aimed at (1) correction of metabolic abnormalities, (2) opening the ductus arteriosus and attempting to improve systemic perfusion with prostaglandin E\textsubscript{1} infusion,\textsuperscript{7} and (3) omitting cardiac catheterization and thereby avoiding the potentially deleterious effects of catheter manipulation and angiography.\textsuperscript{5,8}

This case supports the concept that earlier diagnosis may be facilitated by prenatal recognition of a potentially treatable defect.

The natural history of rapid deterioration postnatally in the neonate with severe ventricular dysfunction and aortic stenosis was interrupted in this case by early diagnosis. The usual mortality for neonates presenting with critical aortic stenosis is in the range of 30% to 50%. We speculated that foreknowledge of the lesion and delivery in a properly equipped tertiary center might decrease this to 20%. In this case we had prospectively estimated that the incremental mortality due to complications of prematurity and respiratory distress would approximately be offset by the advantage of early diagnosis. We recommended cesarean section to minimize any further myocardial ischemia that might occur with a vaginal delivery in the face of oligohydramnios due to prolonged rupture of membranes.

Aortic stenosis vs cardiomyopathy. The differentiation between critical aortic stenosis and primary cardiomyopathy prenatally is of great clinical significance. In our experience with two previous cases of endocardial fibroelastosis and dilated cardiomyopathy diagnosed in utero, there was a normal aortic valve, no poststenotic dilation and, in one case, bilateral atrioventricular valve regurgitation (detected by Doppler fetal echocardiography) in the presence of hydrops fetalis.\textsuperscript{9} Both fetuses died in the prepartum or intrapartum period. The presence of poststenotic dilation of the ascending aorta indicates the potential for the left ventricle to generate a pressure gradient across the aortic valve leading to release of energy in the ascending aorta and resulting dilation. Such patients appear to be good candidates for aortic valvotomy regardless of the appearance of the myocardium on ultrasonography.\textsuperscript{5}

Aortic stenosis vs hypoplastic left ventricle. Dilation of the right ventricle is a constant feature of critical aortic stenosis that results from redistribution of flow to the right ventricle and through the ductus arteriosus secondary to severe left ventricular outflow tract obstruction. This redistribution may give the poorly functioning, hypertrophic left ventricle the appearance of relative, or even absolute, hypoplasia, so that quantitation of left ventricular size is important. The criteria of

m/sec, corresponding to a minimum gradient of 56 mm Hg at a time when the umbilical artery catheter systolic pressure was 50 mm Hg (left ventricular pressure at least 106 mm Hg) (figure 2).

Discussion

Surgical results in critical aortic valve stenosis appear to be related to the size of the left ventricular chamber and the preoperative condition.\textsuperscript{6} Most neonates presenting with this lesion are moribund and have severe metabolic acidosis and low cardiac output due to left ventricular outflow tract obstruction and secondary left ventricular dysfunction. Efforts at im-
Latson et al. can be applied to the term and the near-term fetus, but no similar studies of hypoplasia of the left ventricle in premature infants has been done. Simply applying normal criteria for the left ventricular dimension would be extremely hazardous in such cases because survivability is the paramount issue. Our patient’s size was greater than the minimum known to be compatible with survival in our laboratory. The shape of the left ventricle may be helpful in this regard because hypoplastic, irretrievable ventricles assume a globular instead of an ellipsoidal shape and do not extend to the apex of the heart in a four-chamber view. Differentiation from typical hypoplastic left heart syndrome should not present a significant problem because this is usually associated with both mitral and aortic valve atresia.

Optimal management of potentially life-threatening congenital heart disease recognized before birth should include transport of the mother before the onset of labor to a hospital with adequate cardiac and surgical support services. Cesarean section in the management of abnormalities detected by fetal echocardiography should not be applied in most cases. For example, fetal rhythm abnormalities such as premature atrial contractions, or cardiac defects with little potential to cause perinatal problems such as ventricular septal defects, should be managed after vaginal delivery. When there is evidence of compromised organ function, such as myocardial dysfunction indicated by decreased shortening and increased endocardial echogenicity typical of subendocardial ischemia and fibrosis, then minimizing fetal distress by cesarean section may be indicated. The team approach, with consultation of pediatric cardiologists, neonatologists, and cardiac surgeons can aid in management.

If fetal echocardiography is to fulfill its potential, it must be accurate in both the diagnosis of the anatomic defect and the functional assessment of the fetus so that an accurate prognosis and optimal management can be achieved. Aggressive treatment of potentially salvageable fetuses with cardiac defects could improve outcome.

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