Prediction of Therapeutic Strategy and Outcome for Antenatally Diagnosed Pulmonary Atresia/Stenosis With Intact Ventricular Septum

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Background  The therapeutic strategy for pulmonary atresia (PA) and severe pulmonary stenosis (PS) with intact ventricular septum is controversial. Recent improvements in prenatal detection necessitate the identification of predictors of outcome for appropriate counseling and prenatal management planning.

Methods and Results  Echocardiograms of 18 fetuses antenatally diagnosed with PA (n=14) and PS (n=4) were reviewed and the total cardiac dimension (TCD) and tricuspid valve diameter (TVD) were measured. The right ventricular end-diastolic volume (RVEDV) was calculated from the right ventriculogram of the neonatal period by the percentage of the predicted normal value (%RVEDV). There was a positive correlation between TVD/TCD and %RVEDV (p<0.001). As the initial treatment, balloon atrioseptostomy was performed in 13 cases of TVD/TCD <0.26. As the final treatment, patients with TVD/TCD <0.17 underwent or were planned for the Fontan procedure. Patients with TVD/TCD >0.21 underwent or were planned for biventricular repair. Patients whose TVD/TCD was between 0.17 and 0.21 underwent or were planned for 1.5 ventricular repairs.

Conclusion  TVD/TCD is a useful index for selecting the postnatal initial treatment for PA/PS and to predict the final status of the fetus. Prenatal detection and prediction of the future status is helpful for family counseling. Furthermore, it will help to decide the postnatal management prenatai.

Key Words: Fetal diagnosis; Fetal echocardiography; Pulmonary atresia; Pulmonary stenosis

Pulmonary atresia (PA) with intact ventricular septum and isolated severe valvular pulmonary stenosis (PS) are rare forms of congenital heart disease, the prognoses for which have improved with the progress of surgical procedures. However, the therapeutic plan varies according to the patient’s clinical condition and in recent years, has been determined by the right ventricular end-diastolic volume (RVEDV), tricuspid valve diameter (TVD), the presence of right ventricle-dependent coronary circulation, and the right ventricular configuration. In particular, RVEDV is proposed as an important factor in the therapeutic plan. Because ductus-dependent-type congenital heart diseases such as PA or PS require some interventional treatment soon after birth, the therapeutic plan beyond the fetal period may be particularly important. Recently, diagnostic reports of fetal PA/PS have increased and some of them predict the postnatal prognosis in the fetal period using the TVD and its Z score; however, not all institutes have original Z values. Our aim in this study was to establish an easier factor to use in formulating the therapeutic plan prenatally and for assisting with family counseling.

Methods

Patients

We identified 18 cases of PA and PS diagnosed in utero between 1993 and 2005 at Osaka Medical Center and Research Institute for Maternal and Child Health (Izumi, Japan). Prenatal and postnatal medical records were reviewed for clinical information, including perinatal outcome.

Echocardiography

Fetal echocardiography with cardiovascular anatomical
assessment and Doppler interrogation was performed according to previously described techniques\(^1\)\(^2\)\(^,\)\(^1\)\(^1\) using a variety of Philips HDI (Bothell, WA, USA) systems. All images were recorded on videotape for offline analysis.

Total cardiac dimension (TCD) and TVD were measured from a 4-chamber view of the fetal echocardiogram immediately before closure of the atrioventricular valves and TCD/TVD was calculated (Fig 1).

Angiography
Cardiac catherization and angiography were performed at 0–7 days after birth (mean, 2.2 days), except for 2 low-birth-weight babies. After routine measurements of pressure and oxygen saturation, biplane right ventriculography was performed, except for 2 patients with RVs that were too small for imaging.

The RVEDV was calculated by the area–length method on the largest posteroanterior and lateral projections, and was revised by a regression equation of Shimazaki et al.\(^1\)\(^2\)\(^,\)\(^1\)\(^2\) RVEDV was corrected by normal valve diameter \(^1\)\(^3\) TVD was measured as the maximum negative valve shadow on the largest posteroanterior or lateral projections at early diastole and was corrected by normal valve diameter (TVD).\(^1\)\(^3\)

Statistical Analysis
The echocardiographic and angiographic data measurements are expressed as the mean±standard deviation. All analyses were performed using StatView Ver. 5.0 software (SAS Institute Inc, Cary, NC, USA) Student’s t-test was used to compare the mean values of 2 groups. The relationship between both indices was assessed by linear regression analysis. Statistical significance was taken at p<0.05.

Results
Clinical Characteristics, Associated Lesions and Outcome
Clinical data are summarized in Table 1. Eighteen fetuses were diagnosed antenatally: 14 had PA, and 4 had PS. Gestational age at diagnosis ranged from 27 to 39 weeks (mean, 33.8±3.9 weeks). In all cases, the postnatal diagnosis was the same as the prenatal diagnosis. No patients had prenatal cardiac interventional treatment. No intrauterine, neonatal or infantile deaths occurred.

Three patients had suspected presence of right-ventricle-dependent coronary circulation in the fetal period and postnatal right ventriculography gave the same diagnosis in all cases. In another case, right-ventricle-dependent coronary circulation became clear on postnatal ventriculography.

As the initial treatment in the neonatal period, we performed balloon atrioseptostomy in 13 cases, and balloon pulmonary valvoplasty or Brock operation in the other 5 cases. A low-birth-weight baby of 1.684 g with PS underwent the Brock operation. In addition, we routinely perform balloon atrioseptostomy for Fontan candidates.

As the final treatment, 1 patient underwent the Fontan operation, 3 underwent 1.5 ventricular repairs, and 8 underwent biventricular repair. Five patients were planned for the Fontan operation and 1 for biventricular repair.

No patients died during the observation period of 0–16 years (mean, 4.1±3.6 years). No case had associated extra-cardiac major abnormalities, including chromosomal abnormalities.

Echocardiographic and Angiographic Findings
The echocardiographic data obtained from the fetuses and angiographic data obtained from the neonates are shown in Fig 2. Two cases with very small right ventricles were unable to undergo right ventriculography. In the other 16 cases, there was a positive correlation between TVD/TCD and %RVEDV (p<0.001). TVD/TCD also had a positive correlation with %TVD (r=0.64), but %RVEDV had a stronger correlation (r=0.91).

Initial Treatment and Fetal Echocardiography
The initial treatment is shown in Fig 3. In 11 patients with a TVD/TCD <0.26, including 2 patients with right ventricle-dependent coronary circulation, we chose balloon...
atrioventricular septostomy as the initial treatment, but 4 of the 11 patients underwent biventricular repair as the final treatment, even though we had performed the balloon atrioventricular septostomy during postnatal angiography. As a result, balloon atrioventricular septostomy was not necessary for cases with a TVD/TCD >0.21.

In 4 patients with a TVD/TCD >0.30, we chose balloon pulmonary valvoplasty or the Brock operation.

Final Treatment and Fetal Echocardiography
The final treatment and TVD/TCD are shown in Fig 3. Patients with a TVD/TCD <0.17 underwent or were planned for the Fontan procedure. Patients with a TVD/TCD ranging from 0.17 to 0.21 underwent or were planned for 1.5 ventricular repairs. Patients with a TVD/TCD >0.21 underwent or were planned for biventricular repair. Patients who had successful biventricular repair in the neonatal period did not require further operations.

Discussion
Prenatal Diagnosis of PA and PS
Prenatal echocardiography can accurately diagnose congenital heart disease, including PA and PS. Maeno et al reported that current high resolution ultrasound provides accurate information for prenatal diagnosis of right ventricular outflow obstruction and ventriculocoronary connection. In the present study, all 18 patients had correct fetal diagnoses confirmed by postnatal echocardiography and angiographic examination. With regard to ventriculocoronary connection, we diagnosed 3 cases of right-ventricle-dependent coronary circulation. Although it is important to know whether the coronary artery is interrupted, in these cases it could not be diagnosed. However, we were able to diagnose countercurrent blood flow in the coronary artery and closely monitor it after birth. The 18 patients are still alive, whereas of another 19 patients urgently admitted during the same period to the same hospital after birth without a fetal diagnosis, 2 died, in 1 case because of right-ventricle-dependent coronary circulation. We consider that a prenatal diagnosis of right-ventricle-dependent coronary circulation may improve the prognosis.

Initial Treatment for Fetal PA or PS
We wanted to find an easier factor to use in formulating the prenatal therapeutic plan. The Fontan procedure, 1.5 repairs, and biventricular repair, including balloon valvoplasty, were performed as the surgical treatments of PA and PS. Because there is a difference in right ventricular development, the surgical procedure varies, so the necessary initial treatment at an early age depends on the therapeutic plan. Furthermore, because PA and PS are ductus-dependent-type congenital heart diseases, several initial treatments are required soon after birth, so it is important to devise a therapeutic plan in the fetal period. Angiography and echocardiography are useful for determining the treatment plan. In particular, RVEDV and TVD have been proposed as important factors in the therapeutic plan. In some reports, TVD and its Z score are used to predict postnatal prognosis in the fetal period, but not all institutes have an original Z value. The TVD of the fetal period depends on the size of fetus, so we think that a standardized TVD/TCD is more useful, and our study showed a positive correlation between TVD/TCD and %RVEDV, and between TVD/TCD and %TVD.
Balloon atrioseptostomy is an important initial treatment for PA and PS. We perform it as the initial treatment for Fontan candidates, but in the present study 4 cases underwent biventricular repair as the final treatment, even though we performed balloon atrioseptostomy according to the postnatal angiography. It is difficult to predict the postnatal right ventricular development; however, balloon atrioseptostomy was not necessary for cases of TVD/TCD >0.21 in our clinical data. If no large change occurs in late pregnancy, we can determine the initial treatment prenatally. TVD/TCD in late pregnancy is useful to predict the initial treatment of PA and PS.

Predictors of Outcome in Fetal PA or PS

Another aim of this study was to identify a fetal factor that can predict the outcome of PA and PS. We can predict the final status of the fetus using echocardiography and TVD/TCD was useful for predicting the final status of PA and PS before birth. Three patients had a TVD/TCD between 0.21 and 0.26, and had an unusual clinical course. Although we performed balloon atrioseptostomy as the initial treatment, they underwent biventricular repair as the final treatment. The gradual development of right ventricular volume led us to change the definitive repair from Fontan or 1.5 repair to biventricular repair. Fig 4 shows the postnatal right ventricle and tricuspid valve development. %RVEDV increases with age, but did not occur in patients who underwent the Fontan repair. On the other hand, %TVD was not influenced by growth. Patients with a %TVD >70% underwent biventricular repair or 1.5 repair as the final treatment, whereas patients with a %TVD <70% underwent the Fontan procedure. Some patients with a %TVD >70% gained so much RV volume, but patients with a %TVD <70% did not. RV development may be affected by the postnatal treatment, such as pulmonary insufficiency after the Brock operation or right ventricular outflow tract reconstruction; however, tricuspid valve development was not affected. Thus, %TVD is a useful postnatal index for planning the treatment and for that reason, we think that TVD/TCD is also useful for prediction in the prenatal period, but it is necessary to understand the conditions for such development.

Prenatal detection of this disease and prediction of the future status using this index may help not only postnatal management, but also family counseling. We can explain to the family the strategy and prognosis of the fetus using the index and table. Prenatal detection of this constellation of abnormalities helps in both family counseling and the decisions for postnatal management.

Study Limitations

First, the cases that we reviewed were limited to late pregnancy. We performed fetal echocardiography more than twice for 6 of 18 cases. However, 5 of them were Fontan candidates whose TVD/TCD was very small and did not change significantly in the fetal period, but it is unknown whether TVD/TCD is truly constant. More examinations in the 2nd trimester of pregnancy are necessary.

Second, instrumentation methods for right ventricular volume have not been elucidated. When the right ventricle is very small, there is an aberration in the right ventricular configuration itself, and contrast media does not sufficiently stain the small right ventricle. TVD/TCD should be compared with other methods such as echocardiography.

References


