Prenatal Diagnosis of Double Outlet Right Ventricle Using Advanced Dynamic Flow

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Prenatal diagnosis of double outlet right ventricle remains an ultrasonographic challenge. Although there is a high degree of accuracy in detecting abnormalities in the fetal heart, defining the spatial relationship of the great arteries remains difficult. In this report, we describe a case of double outlet right ventricle with subpulmonary ventricular septal defect and malposition of the great arteries that was readily diagnosed using Advanced Dynamic Flow.

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KEY WORDS: • double outlet right ventricle • prenatal diagnosis
• Dynamic Flow

INTRODUCTION

Double outlet right ventricle (DORV) was first described as early as 1793 [1]. However, its accurate prenatal diagnosis remains a challenge to ultrasonographers. The small target structures, the insufficient distance resolution due to the depth of the lesions of interest, the requirement of a high frame rate for the high heart rate of the fetus, and/or the high level of clutter generated by fetal respiratory movements, all combine to make conventional color Doppler unsatisfactory for the accurate prenatal diagnosis of some congenital fetal heart defects. In particular, there are difficulties in distinguishing DORV from other conotruncal abnormalities such as tetralogy of Fallot or transposition of the great arteries with a ventricular septal defect (VSD) [2–6]. Advanced Dynamic Flow (ADF), however, offers a flow velocity mapping method with wide-band Doppler transmission capabilities that include advantages in fetal vascular imaging with minimized overpainting, improved distance resolution, and a higher frame rate than conventional color Doppler.

In this report, we describe a case of DORV with subpulmonary VSD and malposition of the great arteries that was readily diagnosed using ADF in combination with conventional color Doppler.

CASE REPORT

A 32-year-old woman, gravida 3, para 1 (2 spontaneous abortions), was referred to our institution (Department of Perinatology, National Cardiovascular Center, Osaka, Japan) at 34 weeks’ gestation with a fetal heart anomaly found on routine ultrasound. There was no history of infection during the pregnancy and the patient had not taken any teratogenic drugs. Neither she nor her family had a history of diabetes or other heredofamilial disease.
A Toshiba Aplio 80 system (Toshiba Corporation, Tokyo, Japan) with a 3.5 MHz transducer was used for ultrasonographic examination. No gross fetal deformities were seen other than the abnormalities in the fetal heart. The four-chamber view showed no chamber enlargement. The aorta and the pulmonary artery lay parallel to each other with the aorta on the right, and both arose from the right ventricle. These findings were confirmed using ADF analysis, which also revealed a subpulmonary VSD (Fig. 1). A high blood flow velocity in the pulmonary artery suggested the presence of pulmonary stenosis (Fig. 2).

Amniocentesis was carried out for karyotyping; the results were normal. The family was counseled and the delivery planned.

At 39 weeks’ gestation, after maturation of the cervix, labor was induced. A baby girl weighing 3,212 g was delivered. The Apgar score was 8 at both 1 and 5 minutes. There was slight cyanosis with a saturation of 80% on pulse oximetry. Cardiac examination showed an adynamic precordium with grade 2–3/6 systolic murmurs at the left parasternal border with a single second heart sound (S2). Arterial blood gas analysis of umbilical cord blood showed respiratory acidosis with hypoxemia (pH, 7.296; pCO₂, 45.3 mmHg; pO₂, 20.8 mmHg). The heart was of normal size on chest radiograph, with slightly decreased pulmonary vascular markings.

Out in utero findings were confirmed by neonatal echocardiography. A DORV was seen with a large subpulmonary VSD measuring 10.4 mm in diameter; the aorta was on the right and anterior to the pulmonary artery. There was an infundibular and valvular pulmonary stenosis resulting in a maximum velocity of 195 cm/s. An atrial septal defect 10.3 mm in diameter and a bilateral superior vena cava were also found. At the time of writing, the baby was being managed in the intensive care unit and being prepared for surgery.

**DISCUSSION**

DORV is a complex conotruncal abnormality with considerable anatomic variation [2,4]. Controversy still exists regarding its definition [7]. Witham established the first diagnostic criteria in 1957, describing both the aorta and pulmonary artery as arising from the morphologic right ventricle [8]. Some authors prefer to use the less rigid criteria of Lev and Anderson, requiring that one complete arterial trunk and at least half of the other arterial trunk originate in the right ventricle [2].

The use of conventional echocardiography is inadequate for the diagnosis of this condition. Several studies have shown the benefits of using color flow mapping for the evaluation of structural fetal heart defects and have demonstrated its advantage over conventional echocardiography [9]. Although identification of conotruncal abnormalities has been reported with a high degree of accuracy using color
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Doppler alone, difficulties still lie in defining the spatial relationship of the great arteries [2–6]. In this report, the use of ADF contributed to the ease of diagnosis.

The use of ADF overcomes the limitations of the conventional color Doppler method, namely insufficient distance resolution, low frame rate, high levels of clutter, and blood flow overpainting. Although problems of poor signal sensitivity and low frame rate in deep regions were still encountered [10], further improvements have been made in ADF in order to achieve a higher resolution, greater sensitivity, and improved penetration [11]. These features have enabled us to clearly delineate the origin and spatial relationship of the great arteries in our patient.

As the degree and nature of the hemodynamic disturbance will affect the prognosis of any conotruncal anomaly, an accurate prenatal description is important for the counseling of parents, particularly if termination is considered [3].

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REFERENCES