



Ultrasound evaluation of fetal critical aortic stenosis using the left atrium area/cardiac area ratio and the Doppler patterns in the pulmonary veins

Sachi Sukegawa¹ · Yuka Yamamoto¹ · Keisuke Sato² · Satomi Tanaka¹ · Toshitaka Tanaka¹ · Naoki Mitsuhashi¹

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Abstract

In fetal critical aortic stenosis (AS), a double reverse pattern in the pulmonary veins (PVs) is associated with a poor prognosis. We evaluated the hemodynamic changes using PV Doppler and the left atrium area/cardiac area (LA/CA) ratio in a fetus at 28 weeks of gestation with critical AS complicated with hydrops fetalis, polyhydramnios, and cardiac abnormality. A markedly enlarged LA and severe mitral regurgitation with critical AS were detected, with LA/CA ratio = 0.40 and double reverse pattern with forward/reverse velocity time integral ratio (FRVR) = 1.18 on PV Doppler. After amniotic reduction at 31 weeks, the LA/CA ratio decreased (0.24) and the FRVR in PV increased (7.11). Forward flow through the fetal aorta was seen spontaneously, and hydrops fetalis was relieved with LA volume reduction. A male neonate weighing 2171 g was delivered via cesarean section at 36 weeks with an Apgar score of 5 and 6 at 1 and 5 min, respectively. He required atrial septal opening and bilateral pulmonary artery banding after birth, followed by Norwood operation. The double reverse pattern in PVs might be reversible. The change in FRVR in PVs and LA/CA ratio would be helpful in understanding the hemodynamic change in fetal critical AS.

Keywords Critical aortic stenosis · Pulmonary vein · Fetal echocardiography

Introduction

Fetal critical aortic stenosis (AS) is a challenging lesion with a poor prognosis leading to hydrops fetalis and intrauterine fetal death. Although spontaneous resolution is extremely rare, one case of spontaneous resolution of fetal critical AS before birth was found in the literature [1]. Fetal aortic valvuloplasty is one of the options for achieving biventricular repair for fetuses with a higher risk of emerging hypoplastic left heart syndrome (HLHS); otherwise, almost half of these fetuses undergo single ventricle repair [2]. Indeed, the double reverse pattern in pulmonary veins (PVs) is related to a poor prognosis in fetuses with critical AS [3]. We

encountered a case of critical AS with hydrops fetalis; however, substantial spontaneous resolution of the hydrops was observed and the double reverse pattern in PVs, which was believed to be irreversible, normalized. We report this rare case with the fetal hemodynamic changes using PV Doppler pattern and the left atrium area/cardiac area (LA/CA) ratio to understand this condition better.

Case

A 30-year-old woman, gravida 2, para 1, was referred to our institute at 28⁺¹ weeks of gestation for polyhydramnios, hydrops fetalis, and fetal cardiac abnormality. The fetal growth was within the normal range, but the amniotic fluid level was elevated (amniotic fluid index [AFI] = 22 cm). Fetal skin edema and fluid collection in the fetal chest and abdomen were observed. The echogenic aortic valve in the fetus was not moving, and the size was 1.9 mm (z -score = -6.8). A markedly enlarged LA and severe mitral regurgitation with fetal critical AS were detected (Fig. 1a, b). The mitral regurgitation was 158 cm/s at that time. The

✉ Yuka Yamamoto
yuyamamo@juntendo.ac.jp

¹ Department of Obstetrics and Gynecology, Juntendo University Shizuoka Hospital, 1129 Nagaoka, Izunokuni-sh, Shizuoka 479-2211, Japan

² Department of Cardiology, Shizuoka Children's Hospital, Shizuoka, Japan

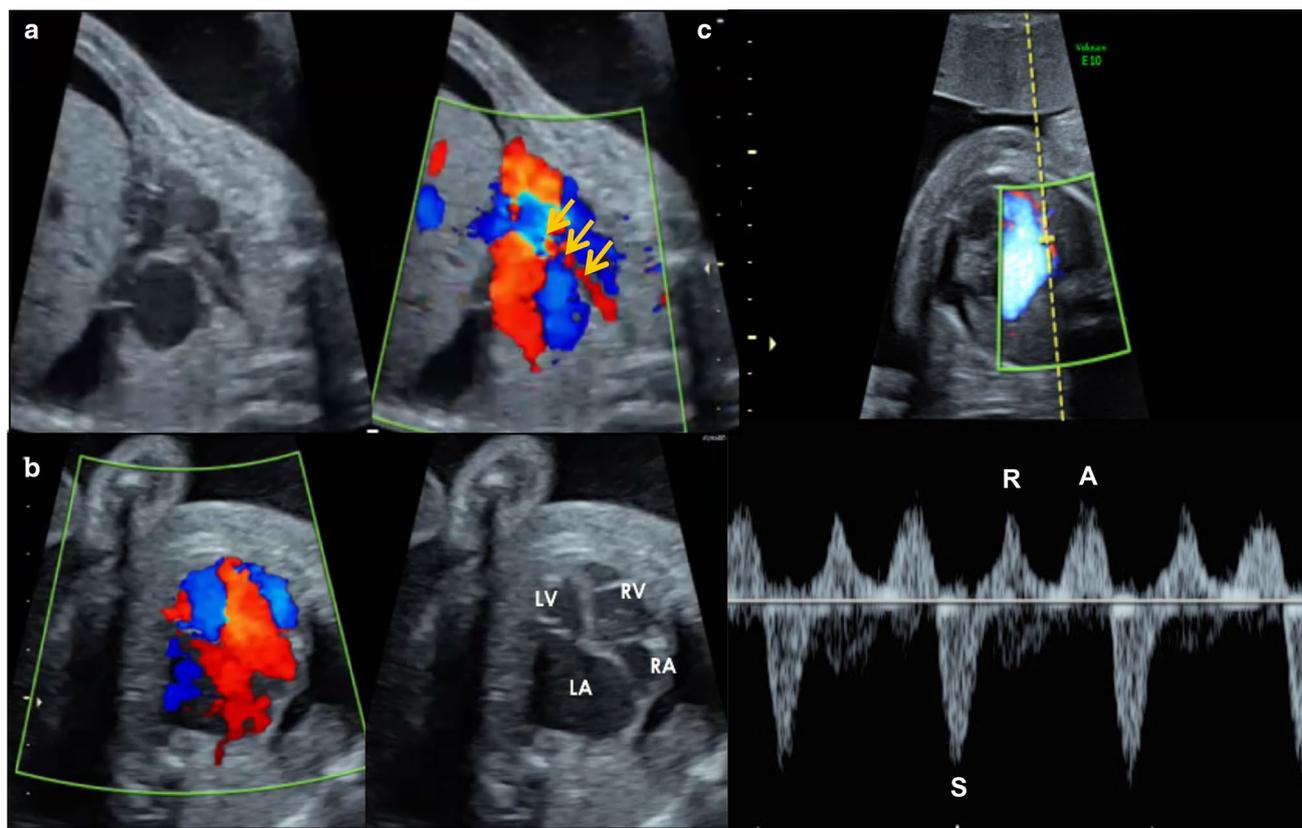


Fig. 1 **a** Sagittal view of the aorta showing forward flow through the aortic valve and retrograde flow of the ascending aorta (yellow arrows). **b** Four-chamber view at 28 weeks showing a markedly enlarged left atrium. *LA* left atrium, *LV* left ventricle, *RA* right

atrium, *RV* right ventricle. **c** Pulsed Doppler of the pulmonary vein showing a double reverse pattern. *S* early systole, *R* first reverse flow, *A* atrial contraction

foramen ovale was bowing to the right with narrowing, and the LA/CA ratio was 0.40. PV Doppler showed a double reverse pattern with a forward/reverse velocity time integral ratio (FRVR) of 1.18 (Fig. 1c). Other Doppler data in the fetus were observed as follows: umbilical arterial (UmA) resistance index (RI)=0.76, middle cerebral arterial (MCA) RI=0.69, and ductus venosus (DV) pulsatility index (PI)=1.08. The Doppler data at 29⁺⁵ weeks of gestation were UmA RI=0.68, MCA RI=0.74, and DV PI=0.94. The FRVR in PV was < 1 and the LA/CA ratio was 0.4 (Fig. 2).

The mother was hospitalized with frequent uterine contractions and discomfort at 30⁺⁵ weeks of gestation due to polyhydramnios. Amniotic fluid reduction was performed to reduce the discomfort at 31⁺¹ weeks of gestation. The forward flow through the fetal aorta was seen spontaneously a few days after amniotic fluid reduction (Fig. 3a). The LA/CA ratio decreased to 0.24 (Fig. 3b), and the PV Doppler pattern showed only a small wave reversal (Fig. 3c) as the FRVR increased to 7.11 (Fig. 2). The hydrops fetalis was relieved with the LA volume reduction, but the left ventricular (LV) function remained poor. The mother was transferred to the

Children's Hospital at 35 weeks of gestation for planned neonatal surgery of the baby. The mother delivered a male neonate weighing 2171 g by cesarean section at 36⁺² weeks of gestation with an Apgar scores of 5 and 6 at 1 and 5 min, respectively. There was endocardial fibroelastosis in the LV, and the motion of the ventricles was insufficient (Fig. 4a, b). The PV Doppler pattern after birth was almost normal (Fig. 4c). The atrial septum was bowing toward the right, and the neonate required atrial septal opening and bilateral pulmonary artery banding right after birth. Subsequently, a Norwood operation was performed, owing to poor LV function.

Discussion

This was a rare case of fetal critical AS with spontaneous resolution of hydrops fetalis that has been reported in only one other patient by Ide et al. [1]. In many cases of fetal critical AS, the prognosis is poor, and some of them result in hydrops fetalis and intrauterine fetal death. With a hypoplastic left heart, it would eventually need a single ventricular

GA	31w1d Amniotic reduction					
	28w1d	29w5d	30w5d	31w5d	32w5d	35w1d
EFBW (g)	1415	1793	1932	1807	2156	2232
AFI (cm)	22	28.4	30.5	31.3	29.3	21.7
Pleural fluid	Bilateral	Bilateral	Right	None	None	None
UmA RI	0.76	0.68	0.80	0.70		0.63
MCA RI	0.69	0.74	0.74	0.74	0.75	0.76
DV PI	1.08	0.94	2.94	0.92	0.71	0.73
CTAR (%)		51.9	56.2	46.9	48.2	42.1
LA/CA ratio	0.4	0.4	0.35	0.24	0.24	0.119
FRVR	1.18	0.79	1.06	7.11	7.87	

Fig. 2 Changes in ultrasound parameters in this case. *GA* gestational age, *EFBW* estimated fetal body weight, *AFI* amniotic fluid index, *UmA* umbilical artery, *RI* resistance index, *MCA* middle cerebral

artery, *DV* ductus venosus, *PI* pulsatility index, *CTAR* cardiothoracic area ratio, *LA* left atrium, *CA* cardiac area, *FRVR* forward/reverse velocity time integral ratio

repair. Fetal aortic valvuloplasty in humans commenced in the early 1990s [4] and leads to biventricular repair in 45% of the patients if successful [5]. To evaluate the availability of fetal therapy, cardiac evaluation of the aorta and LV with z-score is important [6]. In this study, we focused not only on measurements but also on the changes in PV Doppler pattern, because we observed an unusual flow pattern in the PV during the evaluation.

PV Doppler has been reported to be related to left atrial pressure. The PV Doppler pattern has been used for evaluation of the restricted foramen ovale in cases with HLHS. With the increased LA pressure due to the restricted foramen ovale, the forward flow in diastole in the PV decreases, and eventually a to-and-fro pattern in the PV is seen [7]. The to-and-fro flow pattern implies a worse perinatal outcome of the fetus with HLHS. Rogers et al. [3] reported that the double flow reversal pattern in PVs, similar to that of HLHS, would predict the necessity for immediate postnatal intervention in cases with fetal critical AS. According to the report, the double reverse pattern showed that the first wave of reversal (R) occurred during ventricular systole, owing to the mitral reverse, and the second wave of reversal (A)

was associated with atrial contraction. This flow pattern is likely found in patients with the greatest degree of mitral regurgitation and larger LA and higher LA pressure, which is the most severe spectrum of fetal critical AS [3]. In our case, we also observed the double reverse pattern in PV; however, the shape of the PV was not exactly the same as that previously reported (Fig. 1c). It could be attributed to the S wave, which would differ depending on the amount of forward flow into the LA and would be associated with the degree of mitral regurgitation and LA pressure [8]. In our case, there was no flow between the R and A waves, which might have been due to the absence of forward flow through the PV in early diastole for high LA pressure. As shown in a previous study [9], an increased LA pressure induces higher S wave, which was also seen in our case. Quantification of the flow as FRVR might be useful in showing how much flow moves forward into the LA. Although setting the FRVR cutoff for severe critical AS based only on this study is difficult, the FRVR value shows how much less flow is going into the LA. In the case of HLHS, an FRVR in PVs < 5 has been reported as the strongest predictor of the need for emergent atrial septoplasty in the newborn period [10].

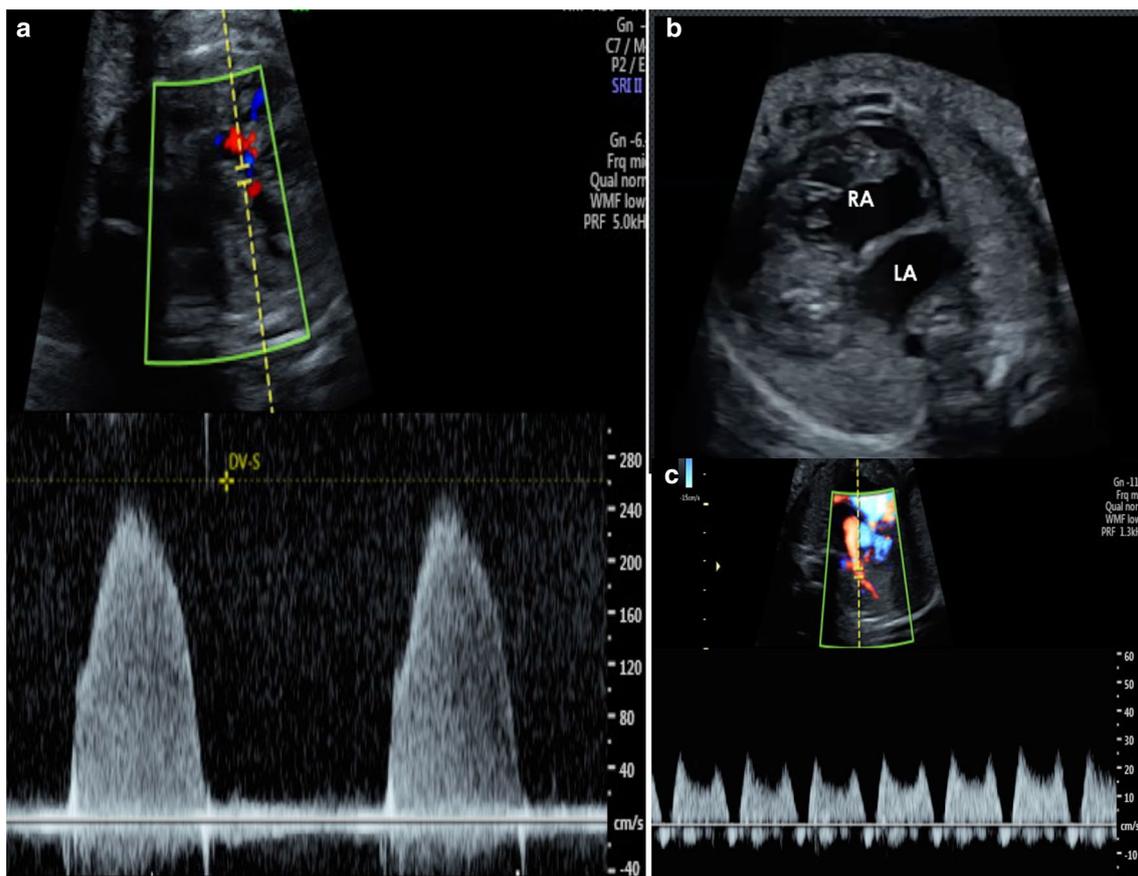


Fig. 3 Ultrasound pictures of the fetus a few days after amniotic fluid reduction: **a** forward flow through the aorta with Doppler, **b** decrease in the size of the LA, **c** pulmonary venous Doppler. LA left atrium, RA right atrium

Furthermore, the double reverse pattern in PV is thought to be a lethal sign in the case of critical AS [3]. Our case showed rare spontaneous resolution of aortic atresia, which led to PV Doppler pattern reversal.

LA size simply reflects the severity of mitral regurgitation. An increase in LA area > 1.5 compared to RA area has been reported to be another lethal sign in fetuses with critical AS [3]. We also observed an obvious decrease in the LA/CA ratio with the resolution of the PV Doppler pattern. A dilated LA with severe mitral regurgitation induces worse fetal circulation, which leads to low cardiac output and hydrops fetalis. Simple observation of the LA is important in understanding the condition in the fetus with critical AS (Fig. 5).

Polyhydramnios was probably caused by the compression from the dilated LA of the fetus. Although the spontaneous resolution of hydrops fetalis after amniotic fluid reduction was an interesting occurrence, it might merely have been a coincidence. In cases of therapeutic amnioreduction in fetuses with twin–twin transfusion syndrome, evidence showed that amnioreduction induced a decrease in afterload of the fetus, which led to improved fetal circulation [11, 12]. A similar phenomenon might have occurred in our case, as well.

In conclusion, we experienced spontaneous resolution of hydrops fetalis with a normalized flow pattern in the PV from a double reverse pattern in a fetus with critical AS. Although this is only one case report, the hemodynamic changes in the fetus with critical AS are interesting.

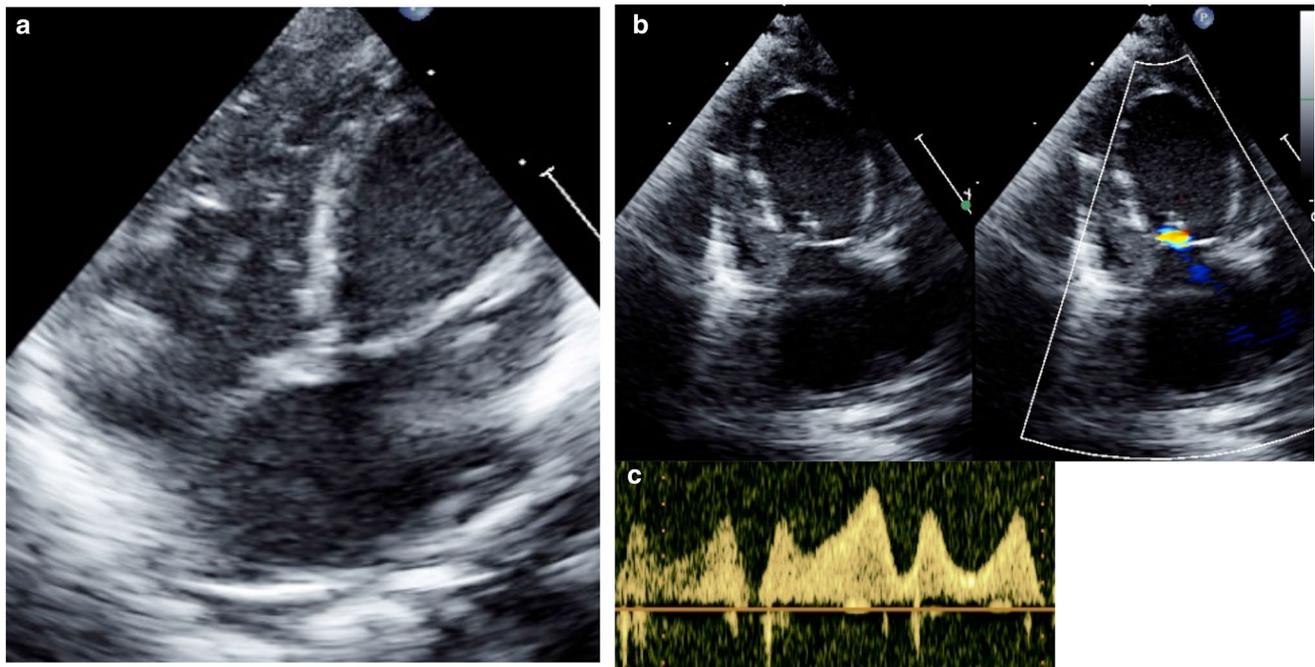


Fig. 4 Ultrasound images after birth. **a** Four-chamber view showing endocardial fibroelastosis in the LV. **b** Mitral valve, with insufficient motion of the ventricles. **c** Almost normal pulmonary venous Doppler pattern

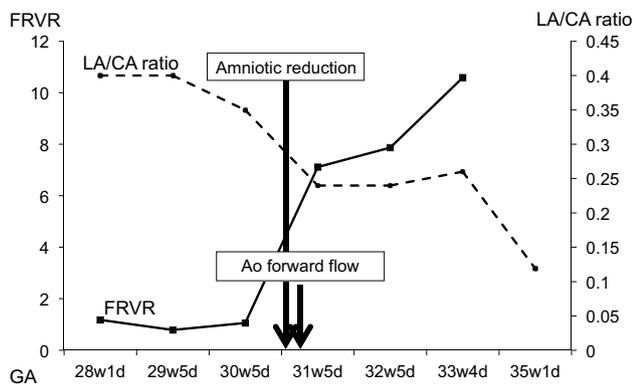


Fig. 5 Clinical course of the pulmonary venous Doppler and the left atrium area/cardiac area ratio. A marked change occurred between 30 and 31 weeks of gestation. When the forward flow through the aorta was seen spontaneously, hydrops fetalis resolved with significant LA volume reduction and increased FRVR. *FRVR* forward/reverse velocity time integral ratio, *LA* left atrium, *CA* cardiac area, *GA* gestational age, *PV* pulmonary vein, *Ao* aorta

Compliance with ethical standards

Conflict of interest Sachi Sukeawa, Yuka Yamamoto, Keisuke Sato, Satomi Tanaka, Toshitaka Tanaka, and Naoki Mitsuhashi declare that they have no conflicts of interest.

Ethical statement All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration

of 1964 and later versions. Informed consent was obtained from the patient for being included in the study.

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