CASE REPORT

Three-dimensional Fetal Echocardiographic Assessment of Persistent Left Superior Vena Cava with Absent Right Superior Vena Cava

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ABSTRACT

We present our experience of diagnosing the fetal persistent left superior vena cava with the absent right superior vena cava using HDlive Flow with spatiotemporal image correlation (STIC). A 28-year-old pregnant Japanese woman, gravida 2, para 1, was referred to us for routine ultrasound screening at 28 weeks and 4 days of gestation. Two-dimensional sonography and radiant flow showed a dilated coronary sinus with the four-chamber view, and revealed a blood vessel on the left side of the pulmonary artery and absence of the superior vena cava with the three-vessel view. However, there was no other cardiac anomaly. Five cardiac views by an HDlive Flow with STIC clearly showed the absence of the right superior vena cava and a persistent left superior vena cava connected to a dilated coronary sinus. HDlive Flow with STIC provides additional diagnostic clues to evaluate abnormal systemic venous return in the fetus.

Keywords: Absent right superior vena cava, Antenatal diagnosis, HDlive Flow, Persistent left superior vena cava, Spatiotemporal image correlation, Three-dimensional fetal echocardiography.

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INTRODUCTION

A persistent left superior vena cava (PLSVC) with an absent right superior vena cava (ARSVC) is an extremely rare condition.¹ The incidence of PLSVC and ARSVC was 0.05% in an autopsy series involving prematurely deceased children, and there were associated cardiac abnormalities in all cases.² However, ARSVC and PLSVC with a structurally normal heart is very rare, and this combination may be noted incidentally in adulthood.¹ Two-dimensional (2D) sonographic diagnostic criteria of PLSVC and ARSVC without any cardiac anomaly are only a markedly enlarged coronary sinus with the four-chamber view and atypical features with the three-vessel view.³ Therefore, cases of this variant may be overlooked in fetuses.

HDlive Flow with spatiotemporal image correlation (STIC) is a new three-dimensional (3D) fetal echocardiography and a useful diagnostic technique for evaluating normal and abnormal fetal cardiac structures, especially normal and abnormal great arteries.⁴⁻⁹ However, to the best of our knowledge, there has been only one report on evaluating abnormal fetal systemic venous return such as PLSVC in the normal fetal heart using HDlive Flow with STIC.¹⁰ In the current study, we present a case of fetal PLSVC and ARSVC without any cardiac abnormality diagnosed with HDlive Flow and STIC.

CASE DESCRIPTION

A 28-year-old pregnant Japanese woman, gravida 2, para 1, was referred to us for our routine ultrasound screening at 28 weeks and 4 days of gestation. Two-dimensional sonography and radiant flow showed a dilated coronary sinus with the four-chamber view, and revealed a blood vessel on the left side of the pulmonary artery and ARSVC with the three-vessel view. However, there was no other cardiac anomaly. Five cardiac views by HDlive Flow with STIC clearly demonstrated ARSVC and PLSVC connected to a dilated coronary sinus (Fig. 1). Five cardiac views of a normal fetal heart at the same age are shown in Figure 2 as a reference.

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At 38 weeks of gestation, she vaginally delivered a viable male infant weighing 3.045 g, with a height of 47.2 cm. The Apgar scores were 8 (1 minute) and 9 (5 minutes), and the umbilical artery blood pH was 7.30. Postnatal echocardiographic examination confirmed the diagnosis. The mother and neonate followed a favorable course after delivery.

DISCUSSION

The incidence of PLSVC detected by prenatal 2D fetal echocardiography was 0.36% in low-risk pregnancies, and 52.3% were isolated anomalies in all fetuses with PLSVC.¹¹ There has been only one report presenting a 3D echocardiographic image

Figs 1A to E: Five cardiac views by HDlive Flow at 28 weeks and 4 days of gestation. Absent right superior vena cava (arrow) and persistent left superior vena cava (PLSVC) connected to the coronary sinus (CS) are clearly depicted. Ao, aorta; AoA, aortic arch; DAo, descending aorta; HV, hepatic vein; IV, innominate vein; IVC, inferior vena cava; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RV, right ventricle. (A) Frontal view; (B) Spatial three-vessel view; (C) Panoramic view; (D) Posterior view; (E) Right lateral view

Figs 2A to E: Five cardiac views of a normal fetal heart at 28 weeks and 4 days of gestation. Ao, aorta; AoA, aortic arch; DA, ductus arteriosus; DAo, descending aorta; HV, hepatic vein; IV, innominate vein; IVC, inferior vena cava; LA, left atrium; LPA, left pulmonary artery; LV, left ventricle; PA, pulmonary artery; PV, pulmonary vessels; RA, right atrium; RPA, right pulmonary artery; RV, right ventricle; SVC, superior vena cava. (A) Frontal view; (B) Spatial three-vessel view; (C) Panoramic view; (D) Posterior view; (E) Right lateral view

of fetal PLSVC without any cardiac anomaly using HDlive Flow with STIC.¹⁰ This technique clearly showed PLSVC on the left side of the pulmonary artery. On the other hand, there have been two reports on the antenatal 2D sonographic diagnosis of PLSVC and ARSVC.^{1,3} The first case was associated with a hypoplastic aortic arch,¹ whereas there were no additional cardiac malformations in the second case.³ However, there has been no study on prenatal diagnosis of PLSVC with ARSVC in the normal heart using 3D fetal echocardiography because this combination is extremely rare. In the current investigation, five 3D cardiac views using HDlive Flow with STIC clearly visualized PLSVC and ARSVC spatially. These cardiac views (frontal, spatial three-vessel, panoramic, posterior, and right lateral views) provided useful diagnostic information to evaluate normal and abnormal fetal cardiac structures.^{7,10,12} In our case, the panoramic view clearly showed PLSVC connected to a dilated coronary sinus, and posterior and right lateral views clearly depicted a characteristic, oval right atrium without the right superior vena cava. This is the first report on 3D ultrasound demonstration of fetal PLSVC with ARSVC *in utero*. HDlive Flow with STIC provides additional diagnostic information to evaluate abnormal systemic venous return in the fetus.

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