ORIGINAL ARTICLE

Analysis of etiology, chromosome and prognosis for small left heart system development in 69 fetuses

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Abstract
Objective: To provide a basis for evaluating the prognosis of small left heart system development in fetuses, we analyzed its related factors.
Methods: The fetal echocardiogram was performed in 3859 pregnant women, and then small left heart system development was identified in 69 fetuses. The data of prenatal and postnatal echocardiograms, postnatal cardiac surgical treatment, chromosome and autopsy after induced labor were analyzed in the 69 fetuses.
Results: Except 1320 cases losing follow-up, 2539 cases had complete data. Among the 2539 cases, small left heart system development was identified in 69 fetuses. Of the 69 fetuses, 12 had hypoplastic left heart syndrome, 20 premature closure of foramen ovale, 13 total anomalous pulmonary venous drainage, 2 common pulmonary vein lumen atresia, 21 aortic coarctation or interruption and 1 right pulmonary hypoplasia. Among the 69 fetuses, chromosome abnormality was found in 7.
Conclusion: There are many etiological factors causing small left heart system development. The prognosis is poor in the fetuses with hypoplastic left heart syndrome, common pulmonary vein lumen atresia, pulmonary hypoplasia, other malformations or/and chromosome abnormality. Fetal echocardiography combined with chromosome examination can provide important bases for making diagnosis and evaluating the prognosis regarding small left heart system development.

Keywords
Chromosome abnormality, echocardiogram, fetus, prenatal diagnosis, small left heart system development

Introduction
Hypoplastic left heart syndrome, a kind of congenital cardiovascular anomaly, is characterized by maldevelopment of ascending aorta, aortic valve, left ventricle or mitral valve; and seriously affects neonatal survival [1–3]. The incidence of hypoplastic left heart syndrome is about 16/100,000 and it accounts for about 1.4% of congenital cardiovascular anomaly. The average survival duration is 4 or 5 d and most of these neonates will die within postnatal 48 h if they do not receive surgical treatment. The neonates with later closed ductus arteriosus may survive for several weeks or months. Surgical treatment is a unique life-saving measure for the neonates with hypoplastic left heart syndrome. With the development of fetal echocardiography, it has been found that besides hypoplastic left heart syndrome, other etiological factors also cause small left heart system development and different etiological factors are associated with prognosis. The purpose of this study was to analyze the etiological factors of small left heart system development and provide a basis for evaluating the prognosis of small left heart system development in fetuses.

Materials and methods
All study methods were approved by the Ethics Committee of Henan Provincial Peoples Hospital. All the subjects enrolled into the study gave formal consent to participate.

Subjects
The prospective study was performed from January 2012 to October 2014. A total of 3859 pregnant women, with a mean age of (29.21 ± 4.68) years (range 16–46) and a mean gestation age of (26.35 ± 3.36) weeks (range 16–40), from Henan Provincial Peoples Hospital, Beijing Anzhen Hospital and the Seventh Peoples Hospital of Zhengzhou underwent fetal echocardiogram. The indications for fetal prenatal
Echocardiography in the study included maternal factors, fetal factors and family factors. The maternal factors referred to the pregnant women who was over 35 years old; had congenital heart disease, infectious diseases, connective tissue disease, endocrine and metabolic diseases, threatened abortion, gestational hypertension, hydramnios, hypannion, positive maternal alloimmune antibody, multiple pregnancy, a history of abnormal pregnancy or a family history of congenital heart disease; or took teratogenic drugs or exposed to teratogenic substances during pregnancy. The fetal factors referred to the fetuses that had cardiac abnormalities showed by conventional ultrasonography, arrhythmia, other organ malformations or chromosomal abnormalities. The family factors referred to that either of parents had congenital heart disease, Mendel disease or birth of fetuses with congenital heart disease in their families. The fetuses with complete data including prenatal and postnatal echocardiograms, postnatal cardiac surgical treatment, chromosome and autopsy after induced labor were enrolled in this study.

Instrument
The fetal echocardiography was performed using GE Voluson E8 ultrasonic instrument with detecting head of RAB4-8, frequency of 4–8 MHz and the mode of Fetal Cardiac, or using Philips iE33 ultrasonic instrument with detecting head of C5–1, frequency of 1–5 MHz and the mode of Fetal Echo. Postnatal echocardiography was performed using Philips iE33 ultrasonic instrument with detecting head of C5-1, frequency of 1–5 MHz and the mode of Pediatrics.

The diagnosis of small left heart system development
According to Standardized Guidelines about Fetal Echocardiography recommended by American Echocardiogram Association [4], we selected four-chamber view, left ventricular outflow view, right ventricular outflow view, three-vessel view, aortic arch view, ductal arch view and caval long-axis view as standard sections. The measurements of hearts were from M Mode tracings and were taken during systole. The reference limits to diagnose small left heart system development are shown in Supplementary Table 1 [5]. The diameters of the left hearts were less than normal values in the same gestational weeks.

Follow-up for small left heart system development
The data including prenatal and postnatal echocardiograms, postnatal cardiac surgical treatment, chromosome and autopsy after induced labor were collected. Prenatal karyotype analysis was performed using fetal amniotic fluid. If the prenatal karyotype analysis failed to be carried out, karyotype analysis was performed using cord blood in the fetuses undergoing induced labor or using venous blood in the fetuses undergoing cardiac surgical treatment.

The reproducibility (intra- and inter-observer) in authors’ hand
The reproducibility (intra- and inter-observer) in authors’ hand was analyzed with Bland–Altman plots for the measurements of the left atrium, left ventricle, right atrium and right ventricle.

Results
Clinical data
In the 3859 pregnant women, their mean age was (29.21 ± 4.68) years (range 16–46) and their mean gestation age was (26.35 ± 3.36) weeks (range 16–40). Of the 3859 cases, 2539 cases (65.8%) had complete data and 1320 cases (34.2%) lost follow-up. Autopsies were performed in the fetuses receiving induced labor and the dead child after birth. Induced labor was performed in 18 cases and 4 cases died after birth (Table 1).

Analysis of etiological factors, chromosome and prognosis for small left heart system development
Among the 2539 cases, the 69 fetuses were first diagnosed with small left heart system development by prenatal echocardiograms, and then the diagnosis of small left heart system development was further confirmed by postnatal cardiac surgery or autopsy. Of the 69 fetuses, 12 had hypoplastic left heart syndrome, 20 premature closure of foramen ovale, 13 total anomalous pulmonary venous drainage, 2 common pulmonary vein lumen atresia, 21 aortic coarctation or interruption and 1 right pulmonary hypoplasia. Among the 69 fetuses, chromosome abnormality was found in 7. The etiological factors, chromosome and prognosis for small left heart system development are shown in Table 1 and Figures 1–6.

Analysis of the reproducibility (intra- and inter-observer) in authors’ hand
Bland–Altman plots indicated that the reproducibility (intra- and inter-observer) in authors’ hand were good in the measurements of the left atrium, left ventricle, right atrium and right ventricle. For the left atrium, 95% confidence interval of intra-observer was (−2.472, 2.142), and 95% confidence interval of inter-observer was (−3.214, 2.434). For the right atrium, 95% confidence interval of intra-observer was (−1.869, 1.589), and 95% confidence interval of inter-observer was (−2.173, 1.683). For the left ventricle, 95% confidence interval of intra-observer was (−2.049, 1.839), and 95% confidence interval of inter-observer was (−2.755, 2.165). For the right ventricle, 95% confidence interval of intra-observer was (−2.572, 1.952), and 95% confidence interval of inter-observer was (−2.622, 2.302) (Figure 7).

Discussion
Fetal heart malformation takes the first place in congenital diseases, and seriously affects fetal intrauterine growth and neonatal survival [6,7]. Fetal echocardiography, one of the most important measures to diagnose fetal heart malformation, can evaluate fetal cardiac structure and function, and has non-invasive and repeatable advantages. With the improvement of ultrasonic instrument, the diagnostic accuracy of the fetal heart malformation gradually increases [8,9].

We should carefully observe whether there is foramen ovale restrictive open, left ventricular fibrosis, pericardial
Table 1. Etiological factors, chromosome and prognosis of small left heart system development.

| Etiological factors                        | Other malformations                                                                 | Cases (n) | Follow-up                                                                                          | Chromosome                        |
|-------------------------------------------|-------------------------------------------------------------------------------------|-----------|---------------------------------------------------------------------------------------------------|-----------------------------------|
| Hypoplastic left heart syndrome           | No                                                                                  | 3         | One-stage Norwood operation was performed after postnatal 3 months and their vital signs are stable at present. | Normal                            |
|                                           | No                                                                                  | 2         | One case died after postnatal 7 d and another case died after postnatal 23 d.                     | Normal                            |
| Central nerve system malformation         |                                                                                     | 4         | Induced labor                                                                                      | One case with 13-trisomy syndrome |
| Digestive system abnormalities            | No                                                                                  | 3         | Induced labor                                                                                      | One case with Turner syndrome     |
|                                           | Moderate or severe hydrothorax or/and ascites                                      | 4         | The 4 cases died after postnatal one hour, 2 h, 7 h and 24 h, respectively                        | Normal                            |
| Premature closure of foramen ovale        |                                                                                     | 16        | Echocardiogram was normal between postnatal 30 and 60 d.                                          | Normal                            |
| Total anomalous pulmonary venous drainage | Pulmonary vein stenosis                                                            | 1         | The case underwent corrective procedure after postnatal 28 d, but died 5 d after operation.       | Cat eye syndrome (Schachenmann syndrome) |
| Common pulmonary vein lumen atresia       | Complete common pulmonary vein lumen atresia combined with single truncus arteriosis, right pulmonary artery stenosis, persistent left superior vena cava and right aortic arch | 1         | Induced labor                                                                                      | Normal                            |
|                                           | Partial common pulmonary vein lumen atresia combined with single ventricle, tricuspid atresia, pulmonary atresia, aortopulmonary collaterals and right aortic arch | 1         | Induced labor                                                                                      | Normal                            |
| Aortic coarctation or interruption        | No                                                                                  | 9         | Successful surgical treatment was performed in all cases between postnatal 3 months and 1 year     | One case with 21-trisomy syndrome  |
|                                           | Ventricular septal defect                                                          | 5         | One case underwent successful end-to-end anastomosis after removal of aortic coarctation, and ventricular septal defect repair. Other cases underwent successful end-to-end anastomosis after removal of aortic coarctation. | One case with 21-trisomy syndrome  |
|                                           | Aortic pulmonic window and the right pulmonary artery originating from the aorta   | 1         | The case underwent successful aortic pulmonic window repair, right pulmonary artery transplantation and corrective procedure with aortic arch after postnatal 4 months. | Normal                            |
|                                           | Urinary system malformation                                                        | 1         | The case underwent end-to-end anastomosis after removal of aortic coarctation on the postnatal 7th day, but died of coagulation disorders 4 d after operation. | Normal                            |
|                                           | Aortic subvalvular stenosis, aortic two leaf malformation, persistent left superior vena cava and reproductive system malformation | 5         | Induced labor                                                                                      | One case with 21-trisomy syndrome, one case with 18-trisomy syndrome and one case with 13-trisomy syndrome |
| Right pulmonary hypoplasia                | No                                                                                  | 1         | Induced labor                                                                                      | Normal                            |
effusion, hydrothorax and ascites when the diagnosis of fetal hypoplastic left heart syndrome is made. In this study, there were 12 fetuses with hypoplastic left heart syndrome. Of the 12 fetuses, 3 without other malformations underwent one-stage Norwood operation after postnatal 3 months and their vital signs were stable after operation; 2 died within postnatal one month; 4 with central nervous system malformation underwent induced labor, of the 4 fetuses, 13-trisomy syndrome was identified in one and other 3 had normal chromosome; 3 with digestive system abnormalities underwent induced labor, but had normal chromosome. We can see from the 12 fetuses that extra-cardiac malformations and chromosome abnormality all can affect the prognosis of hypoplastic left heart syndrome. The staging operations for hypoplastic left heart syndrome include Norwood, hemi-Fontan and Fontan. However, not all fetuses that have undergone one-stage Norwood operation can enter Fontan operation. For the fetuses requiring heart transplantation, 1- and 5-year survival rates of heart transplantation are 61% and 55%; and for the fetuses receiving staging operation, 1- and 5-year survival rates of staging operation are 42% and 38% [10–13].

Premature closure of foramen ovale can lead to right cardiac enlargement and right heart insufficiency, even fetal distress or death. The time and degree of foramen ovale closure is strongly associated with fetal prognosis. Donofrio et al. [14] have reported that the fetuses with premature closure of foramen ovale combined with other cardiac malformations have poor prognosis. Gupta et al. [15] have found that the fetuses only with premature closure of foramen ovale have good prognosis. Uzun et al. [16] have described that if only premature closure of foramen ovale is identified during fetal period, pulmonary hypertension is worthy of notice in neonatal period. In this study, there were 20 fetuses with premature closure of foramen ovale. Of the 20 fetuses, 16 without other malformations and chromosome abnormality had normal echocardiogram between postnatal 30 and 60 d, 4 with moderate or severe hydrothorax or ascites died after postnatal 1 h, 2 h, 7 h and 24 h, respectively. If fetal lungs reach maturity and gestational weeks are more than 36 weeks, early parturition is an approach to save the fetuses with premature closure of foramen ovale. Our study also suggests that the fetuses that only have premature closure of foramen ovale but do not have other malformations, hydrothorax and ascites will have good prognosis. Therefore, in the fetuses with premature closure of foramen ovale, the presence of other malformations, hydrothorax or ascites severely affects fetal prognosis.

Total anomalous pulmonary venous drainage, one of a few diseases requiring emergency operation, accounts for about 1–5% of congenital heart disease. In this study, there

![Figure 1](image1.png)

Figure 1. Hypoplastic left heart syndrome. (a) The four-chamber view of fetal echocardiography shows that the left hearts are markedly smaller than the right ones in 22 gestational weeks. (b) Fetal cardiac autopsy displays that the fetal left hearts are small. RA, right atrium; RV, right ventricle; LA, left atrium; LV, left ventricle.

![Figure 2](image2.png)

Figure 2. Premature closure of foramen ovale combined with massive ascites. (a) The four-chamber view of fetal echocardiography shows that the left hearts are smaller than the right ones in 25 gestational weeks. (b) Abdominal transverse view indicates fetal massive ascites. (c) Fetal cardiac autopsy displays foramen ovale closure. RA, right atrium; RV, right ventricle; LA, left atrium; LV, left ventricle.
were 13 fetuses with total anomalous pulmonary venous drainage. Of the 13 fetuses, 9 without other malformations and chromosome abnormality underwent successful corrective procedure within postnatal 3 months; one with pulmonary vein stenosis and cat eye syndrome died 5 d after operation and 3 with digestive system abnormalities underwent induced labor. With the development of cardiac surgery, the death rate of corrective procedure for total anomalous pulmonary venous drainage have been greatly decreased from 10 to 30% in 1970s to 5–9% at present [17–19]. The degrees of pulmonary venous obstruction and inter-atrial shunt from right to left [20], and the presences of other malformations
and chromosome abnormality all are associated with the prognosis of total anomalous pulmonary venous drainage. Preoperative severe pulmonary venous obstruction, postoperative residuary pulmonary venous obstruction and other malformations all affect the prognosis of total anomalous pulmonary venous drainage.

Common pulmonary vein lumen atresia, one of rate congenital heart malformations, is also the most severe obstructive type of total anomalous pulmonary venous drainage [21]. The fetuses with common pulmonary vein lumen atresia have dyspnea and cyanosis at birth, and will die shortly after birth. The unique outlet of the blood in the lungs is that the blood is drained into the vena systemica through bronchial veins. In this study, two fetuses had common pulmonary vein lumen atresia combined with other cardiac malformations. Of the two fetuses, one had complete common pulmonary vein lumen atresia and another had partial common pulmonary vein lumen atresia. Chromosome abnormality was not found in the two fetuses. The prognosis of common pulmonary vein lumen atresia depends on whether there are collateral veins between the heart atrium and systemic circulation, and other malformations.

Severe aortic coarctation or interruption can cause heart failure and acidosis. If cardiac surgical treatment is not performed in time, about 50% of neonates and about 80% of neonates will die within postnatal 1 month and postnatal 3 months, respectively; and postoperative restenosis may occur in about 50% of neonates [22]. In this study, there were 21 fetuses with aortic coarctation or interruption. Of the 21 fetuses, 9 without other malformations and chromosome abnormality underwent successful corrective procedure between postnatal 3 months and 1 year; 5 had ventricular septal defect, of the 5 fetuses, one had 21-trisomy syndrome; one with urinary system malformation died of coagulation disorders 4 d after corrective procedure; 5 with intra-cardiac or extra-cardiac malformations underwent induced labor, of the 5 fetuses, one had 21-trisomy syndrome, one 18-trisomy syndrome and one 13-trisomy syndrome; one with aortic pulmonic window and the right pulmonary artery originating from the aorta underwent aortic pulmonic window repair, right pulmonary artery transplantation and corrective procedure with aortic arch after postnatal 4 months, and his vital signs were stable after operation. Berry et al. [23] reported that II-type aortic pulmonic window, the right pulmonary artery originating from the aorta, intact ventricular septum, patent ductus arteriosus and aortic arch hypoplasia were named as Berry syndrome; and five neonates with Berry syndrome received corrective procedure, of the five neonates,
Figure 7. Altman–Bland plots showing the intra- and inter-differences of left atrium, right atrium, left ventricle and right ventricle. (a) Intra-observer differences of left atrium. LA, left atrium. (b) Inter-observer differences of left atrium. LA, left atrium. (c) Intra-observer differences of right atrium. RA, right atrium; (d) Inter-observer differences of right atrium. RA, right atrium. (e) Intra-observer differences of left ventricle. LV, left ventricle. (f) Inter-observer differences of left ventricle. LV, left ventricle. (g) Intra-observer differences of right ventricle. RV: right ventricle. (h) Inter-observer differences of right ventricle. RV, right ventricle.
Figure 7. Continued.
Figure 7. Continued.

(a) $\bar{d} \pm SD = -0.105 \pm 0.992 \quad N=20$

(b) $\bar{d} \pm SD = -0.295 \pm 1.255 \quad N=20$
Figure 7. Continued.
operation was successful in two and operation failure occurred in the other three due to the presence of other malformations. Therefore, the presence of other cardiac malformations and chromosome abnormality are strongly associated with the prognosis of aortic coarctation or interruption.

Pulmonary hypoplasia caused by embryonic development disorder, a rare congenital malformation, can occur alone or along with cardiac malformations. The neonates with severe hypoplasia in both lungs will die shortly after birth and pulmonary hypoplasia is one of the causes of perinatal death [24,25]. In this study, one fetus with right pulmonary hypoplasia had no other cardiac malformations, but this pregnant woman decided to terminate the pregnancy.

In summary, there are many etiological factors causing small left heart system development. The prognosis is poor in the fetuses with hypoplastic left heart syndrome, common pulmonary vein lumen atresia, pulmonary hypoplasia, other malformations or/and chromosome abnormality. Fetal echocardiography combined with chromosome examination can provide important bases for making diagnosis and evaluating the prognosis regarding small left heart system development.

Declaration of interest
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