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Neonatal outcomes in fetuses with cardiac anomalies and the impact of delivery route

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Abstract

BACKGROUND—Congenital fetal cardiac anomalies compromise the most common group of fetal structural anomalies. Several previous reports analyzed all types of fetal cardiac anomalies together without individualized neonatal morbidity outcomes based on cardiac defect. Mode of delivery in cases of fetal cardiac anomalies varies greatly as optimal mode of delivery in these complex cases is unknown.

OBJECTIVE—We sought to determine rates of neonatal outcomes for fetal cardiac anomalies and examine the role of attempted route of delivery on neonatal morbidity.

STUDY DESIGN—Gravidas with fetal cardiac anomalies and delivery >34 weeks, excluding stillbirths and aneuploidies (n = 2166 neonates, n = 2701 cardiac anomalies), were analyzed from the Consortium on Safe Labor, a retrospective cohort study of electronic medical records. Cardiac anomalies were determined using *International Classification of Diseases, Ninth Revision* codes and organized based on morphology. Neonates were assigned to each cardiac anomaly classification based on the most severe cardiac defect present. Neonatal outcomes were determined for each fetal cardiac anomaly. Composite neonatal morbidity (serious respiratory morbidity, sepsis, birth trauma, hypoxic ischemic encephalopathy, and neonatal death) was compared between attempted vaginal delivery and planned cesarean delivery for prenatal and postnatal diagnosis. We used multivariate logistic regression to calculate adjusted odds ratio for composite neonatal morbidity controlling for race, parity, body mass index, insurance, gestational age, maternal disease, single or multiple anomalies, and maternal drug use.

RESULTS—Most cardiac anomalies were diagnosed postnatally except hypoplastic left heart syndrome, which had a higher prenatal than postnatal detection rate. Neonatal death occurred in

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8.4% of 107 neonates with conotruncal defects. Serious respiratory morbidity occurred in 54.2% of 83 neonates with left ventricular outflow tract defects. Overall, 76.3% of pregnancies with fetal cardiac anomalies underwent attempted vaginal delivery. Among patients who underwent attempted vaginal delivery, 66.1% had a successful vaginal delivery. Women with a fetal cardiac anomaly diagnosed prenatally were more likely to have a planned cesarean delivery than women with a postnatal diagnosis (31.7 vs 22.8%; $P < .001$). Planned cesarean delivery compared to attempted vaginal delivery was not associated with decreased composite neonatal morbidity for all prenatally diagnosed (adjusted odds ratio, 1.67; 95% confidence interval, 0.85–3.30) or postnatally diagnosed (adjusted odds ratio, 0.99; 95% confidence interval, 0.77–1.27) cardiac anomalies.

CONCLUSION—Most fetal cardiac anomalies were diagnosed postnatally and associated with increased rates of neonatal morbidity. Planned cesarean delivery for prenatally diagnosed cardiac anomalies was not associated with less neonatal morbidity.

Keywords

fetal cardiac anomalies; mode of delivery

Introduction

Congenital fetal cardiac anomalies comprise the most common group of fetal structural anomalies with incidence ranging from 8–12 per 1000 live births depending on the timing of diagnosis.^{1,2} An increasing number of congenital cardiac anomalies are being diagnosed prenatally.^{3,4} The increasing prevalence of diabetes and obesity, both associated with fetal cardiac defects, including a population-attributable risk of 4.2% for pregestational diabetes and 8% for prepregnancy obesity, has also contributed to a rise in fetal cardiac anomalies.^{5–7}

There is limited evidence regarding prenatal detection rates of fetal cardiac anomalies and neonatal outcomes, in part due to nonstandardized reporting of cardiac phenotype. Results of sonographic screening programs for fetal cardiac anomalies increased prenatal detection rate from 35.8–59.7% in an international cohort.^{8,9} Previous reports have grouped all types of fetal cardiac anomalies together for analysis with varying degrees of neonatal morbidity.¹⁰ Botto et al¹¹ proposed a classification scheme for fetal cardiac anomalies incorporating cardiac defect type, cardiac complexity, and extracardiac anomalies. Classifying by these categories allows for better evaluation of specific rates of prenatal detection and neonatal morbidity for fetal cardiac anomalies.

Mode of delivery in pregnancies with suspected fetal cardiac anomalies varies greatly.^{12,13} Depending on the severity of the cardiac lesion, the fetus may be unable to tolerate the stress of labor or may need urgent cardiovascular surgery. As the optimal mode of delivery in these complex pregnancies is still unknown, scheduled cesarean delivery (CD) for the indication of fetal cardiac anomalies is being performed.¹² In a retrospective review of infants with fetal cardiac anomalies, prenatal diagnosis decreased the likelihood of spontaneous labor compared to induced labor or planned CD.¹⁴

Our objectives were to determine rates of neonatal outcomes for fetal cardiac anomalies by type of lesion and examine the role of attempted route of delivery on neonatal morbidity using data from a large US obstetrical cohort.

Materials and Methods

We performed a retrospective cohort study on singleton and multiple gestations complicated by fetal cardiac anomalies that resulted in a live birth 34 weeks of gestation using data from the Consortium on Safe Labor, which has been described elsewhere.¹⁵ Stillbirths and aneuploidies were excluded. The consortium included 12 sites (19 hospitals) across the United States with deliveries from 2002 through 2008. Maternal demographic data, labor and delivery information, birth records, and neonatal information were extracted from the patient electronic medical records and supplemented with discharge summary codes. Validation studies comparing discharge diagnoses in the electronic medical record with a manual review confirmed 91.1–99.9% concordance for 20 variables including attempted mode of delivery.¹⁵ Among neonates delivered with cardiac anomalies, 98.3% delivered at an academic institution, 98.0% delivered at a tertiary care center, 98.3% had 24-hour neonatology coverage, and 96.5% had a level-3 neonatal intensive care unit (NICU). Institutional review board approval was obtained from each participating institution within the Consortium on Safe Labor and from our institution for data analysis.

Fetal cardiac anomalies were identified based on *International Classification of Diseases, Ninth Revision (ICD-9)* coding criteria (Appendix A). We determined prenatal diagnosis using antepartum condition *ICD-9* description (0.03 modifier) or the comprehensive *ICD-9* code for fetal anomaly affecting the antepartum period (655.0). Cardiac anomalies were grouped according to the congenital heart disease classification scheme proposed by Botto et al¹¹ to include cardiac defect morphology, complexity, and accompanying extracardiac defects. In cases of multiple fetal cardiac anomalies, the neonate was included in the most severe cardiac anomaly classification. The severity of cardiac defect was determined by a hierarchical list of highest to lowest rate of neonatal mortality.

The rate of cardiac anomalies diagnosed both prenatally and postnatally were determined and compared with rates of prenatal diagnosis reported from the literature. Rates of neonatal death, serious respiratory morbidity, NICU stay > 72 hours, and 5-minute Apgar score <7 were calculated for each type of fetal cardiac anomaly. Serious respiratory morbidity was defined by the need for NICU admission for respiratory support requiring any of the following: oxygen therapy with nasal cannulae, continuous positive airway pressure, bilevel positive airway pressure, ventilator use, diagnosis of respiratory distress syndrome, hyaline membrane disease, pneumonia, apnea, bradycardia, pulmonary hypertension, pneumothorax, meconium aspiration, or pulmonary hypoplasia.

Gestational age at delivery, maternal age, prepregnancy body mass index, race, insurance, parity, CD history in addition to maternal diabetes, smoking, and drug use status were compared between attempted vaginal delivery and planned CD for all classifications of fetal cardiac anomalies. Attempted vaginal delivery was defined as spontaneous labor onset or labor induction. We chose attempted route of delivery instead of actual route of delivery as

women who require CD in labor are more likely to have a higher maternal and neonatal complication rate, which should be analyzed with the attempted delivery group to limit bias.
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We determined composite neonatal morbidity according to gestational age at delivery and attempted route of delivery for each classification of fetal anomaly. To better identify the role of prenatal diagnosis on attempted route of delivery and composite neonatal outcome, we analyzed fetal cardiac anomalies according to prenatal or postnatal diagnosis and attempted route of delivery. Composite neonatal morbidity included serious respiratory morbidity, sepsis, birth trauma, hypoxic ischemic encephalopathy/asphyxia/seizures, and neonatal mortality.

Demographic characteristic comparison along with univariate analyses were performed with χ^2 and Fisher exact test. Cochran-Armitage trend test was performed to compare composite neonatal morbidity across gestational week of delivery. We performed multivariable logistic regression to determine adjusted odds ratio (aOR) for composite neonatal morbidity controlling for maternal race, parity, body mass index, insurance status, gestational age, maternal disease (diabetes, systemic lupus erythematosus, renal disease, HIV, preeclampsia), single or multiple cardiac and extracardiac anomalies, and maternal drug use history for all classifications of fetal cardiac anomalies according to prenatal or postnatal diagnosis and attempted route of delivery. Generalized estimating equations were used to account for correlations between pregnancies to the same mother. A value of $P < .05$ was considered significant. Statistical analysis was performed with software (SAS, Version 9.1; SAS Institute Inc, Cary, NC).

Results

There were 2319 live-born neonates with fetal cardiac anomalies in the database. In all, 153 cases with aneuploidy were excluded. The final cohort included 2166 neonates representing 2701 total fetal cardiac anomalies (Figure). Of 2166 neonates, 69% had an isolated cardiac anomaly, 16.2% had >1 cardiac anomaly, and 14.8% had both a cardiac anomaly and extracardiac anomaly.

Rates of cardiac anomalies grouped according to the classification proposed by Botto et al¹¹ are presented in Table 1. Most cardiac anomalies were diagnosed postnatally except hypoplastic left heart syndrome, which had a higher prenatal than postnatal detection rate (Table 1). Rates of neonatal death, respiratory morbidity, NICU admission >72 hours, and 5-minute Apgar score <7 varied among cardiac anomalies (Table 1). Fetal cardiac anomalies with the highest neonatal mortality rate were double outlet right ventricle and other transposition of the great vessels, both with 25% neonatal mortality. Other specific fetal cardiac anomalies with high neonatal mortality rates included common truncus (20.0%) and hypoplastic left heart syndrome (16.1%). Neonates with double outlet right ventricle and total anomalous pulmonary venous connection had the highest rates of serious respiratory morbidity at >80%.

Overall, 76.3% of 2166 pregnancies with fetal cardiac anomalies underwent attempted vaginal delivery and 23.7% of 2166 underwent planned CD (Appendix B). Among patients who underwent attempted vaginal delivery, 66.1% of 1652 had a successful vaginal delivery. The majority of planned CD, 70.2% of 514, occurred at <39 weeks' gestation. In contrast, 51.6% of 1011 cases of spontaneous labor and 45.6% of 641 labor inductions occurred at <39 weeks' gestation.

We evaluated composite neonatal outcome by gestational age and attempted route of delivery for the most common fetal cardiac anomalies in our cohort (Table 2). Composite neonatal outcome varied by completed weeks of gestation and was highest for most classes of fetal cardiac anomalies <37 6/7 weeks. Planned CD was not associated with a decreased risk of composite neonatal outcome for any type of fetal cardiac anomaly at any of the gestational age ranges included in our cohort.

Women with a fetal cardiac anomaly diagnosed prenatally were more likely to have a planned CD than women with a postnatal diagnosis (31.7 vs 22.8%, $P < .001$). Planned CD compared to attempted vaginal delivery was not associated with decreased composite neonatal outcome for all prenatally diagnosed (aOR, 1.67; 95% confidence interval [CI], 0.85–3.30) as well as postnatally diagnosed (aOR, 0.99; 95% CI, 0.77–1.27) cardiac anomalies.

Comment

The prenatal diagnosis of fetal cardiac anomalies remained challenging as most infants with congenital heart defects were not identified until after birth. Neonatal mortality and morbidity varied with specific fetal cardiac anomaly morphology. Attempted vaginal delivery in cases of fetal cardiac anomalies was associated with a 66.1% successful vaginal delivery rate. Planned CD for prenatally diagnosed fetal cardiac anomalies occurred at a higher rate than in postnatally diagnosed cases, but importantly, planned CD was not associated with a decreased risk of composite neonatal morbidity compared to attempted vaginal delivery.

The rates of neonatal outcomes by cardiac lesion type can help individualize patient counseling on neonatal outcomes. For example, among specific types of conotruncal defects, the neonatal death rate varied from 0% for complete transposition of the great vessels to 25.0% for double outlet right ventricle. This variation is crucial to determining the optimal center for delivery given the level of acuity and for patient counseling. Previous reports have attempted to document neonatal morbidity outcomes for fetal cardiac anomalies.^{17,18} The specific fetal anomalies and primary neonatal outcomes studied were limited and a comprehensive report across all classifications of fetal cardiac anomalies was not provided.

Obstetrician-gynecologists currently practice in an environment where decreased rates of CD are recommended. Investigation into alternatives and policies to decrease CD rates have been proposed by several national organizations.^{19,20} Our findings are consistent with previous studies reporting that planned CD for fetal cardiac anomalies are being performed. Dadlez et al¹² reported a decrease in scheduled CD for pregnancies with fetal cardiac

anomalies after implementation of policies to decrease early term delivery in 2009 (49% vs 23%, $P < .001$). However, fetal cardiac anomalies still served as an indication for planned CD at the end of the study period. In a study using data from the University of California Fetal Consortium, neonates with prenatally diagnosed cardiac anomalies were born earlier (38.1 vs 39 weeks; $P < .001$) and more often by planned CD (35.6 vs 26.2%; $P = .004$) than neonates with postnatal diagnosis.²¹ Our findings are also consistent with previous reports that mode of delivery for fetal cardiac anomalies may not affect neonatal outcome. In a cohort of 329 patients, Trento et al¹⁴ found that prenatal diagnosis of congenital heart disease increased likelihood of planned CD (odds ratio, 2.6; 95% CI, 1.6–4.5; $P < .001$) but had no effect on Apgar score, neonatal intubation, or survival to discharge.

Limitations of our research include that it is a retrospective study from a large obstetric cohort developed using electronic medical records. We also determined cardiac anomaly classification with *ICD-9* codes. One previous report studied the accuracy of *ICD-9* coding for congenital heart disease and found the sensitivity of coding was 83% for tetralogy of Fallot, 100% for transposition of the great vessels, and 95% for hypoplastic left heart syndrome.²² Therefore, our results should be interpreted with caution as evaluation of coding retrospectively may not reflect a complete, accurate diagnoses. We did not have information about the type of physician who performed the coding (obstetrician-gynecologist, maternal-fetal medicine specialist, or neonatologist) available in our database and, therefore, did not account for this in our analysis. We could not stratify differences in outcome between planned CD and attempted vaginal delivery based on fetal cardiac disease severity (presence of accompanying fetal hydrops or fetal growth restriction) as this information was not consistently reported in our database. A post-hoc power calculation demonstrated we had 61% power to detect a difference in composite neonatal morbidity between attempted vaginal delivery and planned CD for prenatally diagnosed fetal cardiac anomalies. Given the rate of fetal cardiac anomalies in the population, amassing a larger sample size is difficult and is a limitation of our study. The strengths of our study include the large number of patients included (2166 neonates corresponding to 2701 fetal cardiac anomalies) and the implementation of a standardized classification scheme to document prenatal detection rates and neonatal outcomes for specific fetal cardiac anomalies.

Our study provides data for a multidisciplinary team of physicians to inform and counsel patients about neonatal outcomes for specific cardiac anomalies. We also successfully utilized a comprehensive classification system to better individualize patient care. We found that planned CD in cases of prenatally diagnosed fetal cardiac anomalies was not associated with decreased composite neonatal morbidity. Scheduled CD for the sole indication of fetal cardiac anomaly may not improve neonatal outcomes. An evaluation of neonatal morbidity by attempted route of delivery stratified by severity of fetal cardiac anomaly is needed to determine the optimal mode of delivery for fetal cardiac anomalies.

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References

1. Hoffman JI, Kaplan S, Liberthson RR. Prevalence of congenital heart disease. *Am Heart J*. 2004; 147:425–39. [PubMed: 14999190]
2. Hoffman JI. The global burden of congenital heart disease. *Cardiovasc J Afr*. 2013; 24:141–5. [PubMed: 24217047]
3. Garne E, Loane M, Dolk H, et al. Prenatal diagnosis of severe structural congenital malformations in Europe. *Ultrasound Obstet Gynecol*. 2005; 25:6–11. [PubMed: 15619321]
4. Garne E, Stoll C, Clementi M. Euroscan Group. Evaluation of prenatal diagnosis of congenital heart diseases by ultrasound: experience from 20 European registries. *Ultrasound Obstet Gynecol*. 2001; 17:386–91. [PubMed: 11380961]
5. Liu S, Rouleau J, Leon JA, Suave R, Joseph KS, Raj JG. Impact of pre-pregnancy diabetes mellitus on congenital anomalies, Canada, 2002–2012. *Health Promot Chronic Dis Prev Can*. 2015; 35:79–84. [PubMed: 26186019]
6. Brite J, Laughon SK, Troendle J, Mills J. Maternal overweight and obesity and risk of congenital heart defects in offspring. *Int J Obes*. 2014; 38:878–82.
7. Simeone R, Tinker SC, Gilboa M, et al. Proportion of selected congenital heart defects attributable to recognized risk factors. *Ann Epidemiol*. 2016; 26:838–45. [PubMed: 27894567]
8. van Velzen CL, Clur SA, Rijlaarsdam MEB, et al. Prenatal detection of congenital heart disease—results of a national screening program. *BJOG*. 2016; 123:400–7. [PubMed: 25625301]
9. Rossier MC, Mivelaz Y, Addor MC, Sekarski N, Meijboom EJ, Vial Y. Evaluation of prenatal diagnosis of congenital heart disease in regional controlled case study. *Swiss Med Wkly*. 2014; 144:w14068. [PubMed: 25474330]
10. Almeida LF, Araujo E Júnior, Crott GC, et al. Epidemiological risk factors and perinatal outcomes of congenital anomalies. *Rev Bras Ginecol Obstet*. 2016; 38:348–55. [PubMed: 27459392]
11. Botto LD, Lin AE, Riehle-Colarusso T, Malik S, Correa A. National Birth Defects Prevention Study. Seeking causes: classifying and evaluating congenital heart defects in etiologic studies. *Birth Defects Res A Clin Mol Teratol*. 2007; 79:714–27. [PubMed: 17729292]
12. Dadlez NM, Brubaker SG, Simpson LL, Yilmaz B, Williams IA. Impact of change in delivery practice on neonatal and maternal outcomes in cases of significant congenital heart disease. *Congenit Heart Dis*. 2014; 9:368–7. [PubMed: 25371936]
13. Reis PM, Punch MR, Bove EL, van de Ven CJ. Obstetric management of 219 infants with hypoplastic left heart syndrome. *Am J Obstet Gynecol*. 1998; 179:1150–4. [PubMed: 9822492]
14. Trento LU, Pruetz JD, Chang RK, Dettlerich J, Sklansky MS. Prenatal diagnosis of congenital heart disease: impact of mode of delivery on neonatal outcome. *Prenat Diagn*. 2012; 32:1250–5. [PubMed: 23080120]
15. Zhang J, Troendle J, Reddy UM, et al. Contemporary cesarean delivery practice in the United States. *Am J Obstet Gynecol*. 2010; 203:326e1–10. [PubMed: 20708166]
16. Reddy UM, Zhang J, Sun L, Chen Z, Raju TN, Laughon SK. Neonatal mortality by attempted route of delivery in early preterm birth. *Am J Obstet Gynecol*. 2012; 207:117e1–8. [PubMed: 22840720]

17. Freud LR, Escobar-Diaz MC, Kalish BT, et al. Outcomes and predictors of perinatal mortality in fetuses with Ebstein anomaly or tricuspid valve dysplasia in the current era: a multicenter study. *Circulation*. 2015; 132:481–9. [PubMed: 26059011]
18. Holland BJ, Myers JA, Woods CR. Prenatal diagnosis or critical congenital heart disease reduces risk of death from cardiovascular compromise prior to planned neonatal cardiac surgery: a meta-analysis. *Ultrasound Obstet Gynecol*. 2015; 45:631–8. [PubMed: 25904437]
19. Healthy People 2020. [Accessed Oct. 20, 2016] Maternal Infant and Child Health. MICH 7.1 Reduce cesarean births among low-risk women with no prior cesarean births. Available at: <https://www.healthypeople.gov/2020/data-search/Search-the-Data#srch=cesarean>
20. American College of Obstetricians and Gynecologists. Safe prevention of the primary cesarean delivery. Obstetric care consensus no 1. *Obstet Gynecol*. 2014; 123:693–711. [PubMed: 24553167]
21. Peyvandi S, Nguyen TA, Almeida-Jones M, et al. Timing and mode of delivery in prenatally diagnosed heart disease—an analysis of practices within the University of California Fetal Consortium (UCfC). *Pediatr Cardiol*. 2017; 38:588–95. [PubMed: 28078382]
22. Strickland MJ, Riehle-Colarusso TJ, Jacobs JP, et al. The importance of nomenclature for congenital cardiac disease: implications for research and evaluation. *Cardiol Young*. 2008; 18:92–100. [PubMed: 19063779]

APPENDIX A. Fetal cardiac anomaly classification with corresponding International Classification of Diseases, Ninth Revision code

Hierarchy of inclusion ^a	ICD-9 code	Description
1. Conotruncal defects		
1a.	745.11	Double outlet right ventricle
1b.	745.0	Common truncus
1c.	745.19	Other transposition of great vessels
1d.	745.2	Tetralogy of Fallot
1e.	746.09	Other congenital anomalies of pulmonary valve
1f.	745.10	Complete transposition of great vessels
2. LVOTO defects		
2a.	746.7	Hypoplastic left heart syndrome
2b.	747.10	Coarctation of aorta
2c.	746.3	Congenital stenosis of aortic valve
3. RVOTO defects		
3a.	746.01	Atresia of pulmonary valve, congenital
3b.	746.2	Ebstein anomaly
3c.	746.02	Stenosis of pulmonary valve
4. Congenital anomalies pulmonary artery		
4a.	747.41	Total anomalous pulmonary venous congestion
4b.	747.3	Anomalies pulmonary artery
5. Atrial septal defects		
5.	745.5	Atrial septal defects
6. Ventricular septal defects		
6.	745.4	Ventricular septal defects

Hierarchy of inclusion ^a	ICD-9 code	Description
7. Heterotaxy		
7.	759.3	Heterotaxy
8. Complex defects		
8a.	745.3	Common ventricle
8b.	745.12	Corrected transposition of great vessels
9. Atrioventricular septal defects		
9a.	745.60	Endocardial cushion defect, unspecified type
9b.	745.61	Other endocardial cushion defects
9c.	745.69	Ostium primum defect

ICD-9, *International Classification of Diseases, Ninth Revision*; LVOTO, left ventricular outflow tract obstruction; RVOTO, right ventricular outflow tract obstruction.

^aBased on defect severity as measured by neonatal mortality.

APPENDIX B. Demographic characteristics of pregnancies with congenital fetal cardiac anomalies according to attempted route of delivery

	Attempted vaginal delivery N = 1652 ^a			P value
	Spontaneous labor N = 1011	Labor induction N = 641	Planned CD N = 514	
Prenatal diagnosis of fetal cardiac anomaly	60 (5.9)	97 (15.1)	73 (14.2)	<.01
Mode of delivery				
CD	333 (32.9)	227 (35.4)	514 (100)	<.01
Vaginal	678 (67.1)	414 (64.6)	0 (0)	
Gestational age, wk				
34 0/7–36 6/7	229 (22.7)	102 (15.9)	121 (23.5)	<.01
37 0/7–37 6/7	117 (11.6)	85 (13.3)	95 (18.5)	
38 0/7–38 6/7	176 (17.4)	105 (16.4)	145 (28.2)	
39 0/7–39 6/7	254 (25.1)	190 (29.6)	106 (20.6)	
40 0/7–40 6/7	161 (15.9)	96 (15.0)	36 (7.0)	
41	74 (7.3)	63 (9.8)	11 (2.1)	
Maternal age, y				
<18	37 (3.7)	25 (3.9)	7 (1.4)	<.01
18–34	847 (83.8)	515 (80.5)	372 (72.7)	
>35	127 (12.6)	100 (15.6)	133 (26.0)	
Prepregnancy body mass index, kg/m ²				
<25	375 (37.1)	227 (35.4)	119 (23.2)	<.01
25–29.9	163 (16.1)	97 (15.1)	85 (16.5)	
>30	113 (11.2)	103 (16.1)	97 (18.9)	
Missing	360 (35.6)	214 (33.4)	213 (41.4)	

	Attempted vaginal delivery N = 1652 ^a			P value
	Spontaneous labor N = 1011	Labor induction N = 641	Planned CD N = 514	
Race				
Caucasian	546 (54.0)	310 (48.4)	186 (36.2)	<.01
African American	158 (15.6)	140 (21.8)	149 (29.0)	
Hispanic	171 (16.9)	126 (19.7)	114 (22.2)	
Asian or Pacific Islander	56 (5.5)	27 (4.2)	23 (4.5)	
Other	80 (7.9)	38 (5.9)	42 (8.2)	
Insurance type				
Private	612 (60.5)	333 (52.0)	252 (49.0)	<.01
Public	330 (32.6)	220 (34.3)	208 (40.5)	
Self-pay	12 (1.2)	4 (0.6)	4 (0.8)	
Other	57 (5.6)	84 (13.1)	50 (9.7)	
Parity				
Nulliparous	373 (36.9)	321 (50.1)	145 (28.4)	<.01
1	278 (27.5)	175 (27.3)	208 (40.5)	
>2	360 (35.6)	145 (22.6)	161 (31.3)	
History of CD				
Yes	160 (15.8)	19 (3.0)	296 (57.6)	<.01
No/unknown	851 (84.2)	622 (97.0)	218 (42.4)	
Smoker				
Yes	72 (7.1)	52 (8.1)	33 (6.4)	.53
No/unknown	939 (92.9)	589 (91.9)	481 (93.6)	
Alcohol/drug use				
Yes	45 (4.5)	25 (3.9)	16 (3.1)	.45
No/unknown	966 (95.5)	616 (96.1)	498 (96.9)	
Diabetes				
Yes	87 (8.6)	100 (15.6)	123 (23.9)	<.01
No/unknown	924 (91.4)	541 (84.4)	394 (76.1)	

Data presented as n (%) unless otherwise specified.

CD, cesarean delivery.

^aColumn heading N corresponds to 2166 total neonates included in analysis with stillbirths and chromosomal anomalies excluded.

FIGURE. Flow diagram of included and excluded patients

*Individual patients may have >1 fetal cardiac anomaly so total number of anomalies is greater than total number of patients included in analysis.

TABLE 1
Most common fetal cardiac anomaly classifications with rates of prenatal diagnosis and neonatal outcomes

Description	Fetal anomaly with prenatal diagnosis, n/N (%)	Fetal anomaly with postnatal diagnosis, n/N (%)	Prenatal detection available in current literature, ^a %	Neonatal death, n/N (%)	Serious respiratory morbidity, n/N (%)	NICU admission >72 h, n (%) ^b	5-min Apgar score <7, n (%) ^b
Conotruncal defects, N = 107 ^c	34 (31.8)	73 (68.2)		9 (8.4)	54 (50.5)	45 (45.0)	7 (6.7)
Double outlet right ventricle, N = 24	12 (50.0)	12 (50.0)	68.6–82.9	6 (25.0)	20 (83.3)	16 (72.7)	4 (17.4)
Common truncus, N = 10	5 (50.0)	5 (50.0)	68.6–85.2	2 (20.0)	3 (30.0)	4 (44.4)	2 (20.0)
Other transposition of great vessels, N = 4	0 (0)	4 (100)	NA	1 (25.0)	2 (50.0)	2 (50.0)	0 (0)
Tetralogy of Fallot, N = 49	11 (22.5)	38 (77.6)	41.7–68.6	0 (0)	15 (30.6)	17 (35.4)	1 (2.0)
Other congenital anomalies of pulmonary valve, N = 3	1 (33.3)	2 (66.7)	NA	0 (0)	2 (66.7)	1 (50.0)	0 (0)
Complete transposition of great vessels, N = 17	5 (29.4)	12 (70.6)	44.2–66.0	0 (0)	12 (70.6)	5 (33.3)	0 (0)
LVOTO defects, N = 83	34 (41.0)	49 (59.0)		5 (6.0)	45 (54.2)	29 (37.7)	2 (2.4)
Hypoplastic left heart syndrome, N = 31	20 (64.5)	11 (35.5)	87.5–97.6	5 (16.1)	19 (61.3)	6 (20.7)	0 (0)
Coarctation of aorta, N = 36	9 (25.0)	27 (75.0)	25.7–25.9	0 (0)	18 (50.0)	16 (50.0)	2 (5.6)
Congenital stenosis of aortic valve, N = 16	5 (31.3)	11 (68.8)	25.9–26.9	0 (0)	8 (50.0)	7 (43.8)	0 (0)
RVOTO defects, N = 72	13 (18.1)	26 (81.9)		2 (2.8)	29 (40.3)	30 (46.2)	5 (6.9)
Atresia of pulmonary valve, congenital, N = 8	4 (50.0)	4 (50.0)	61.9–68.6	1 (12.5)	6 (75.0)	4 (66.7)	1 (12.5)
Ebstein anomaly, N = 9	4 (44.4)	5 (55.6)	25.9	1 (11.1)	6 (66.7)	7 (77.8)	1 (11.1)
Stenosis of pulmonary valve, congenital, N = 55	5 (9.1)	50 (90.9)	19.2–25.9	0 (0)	17 (30.9)	19 (38.0)	3 (5.5)
Congenital anomalies pulmonary artery, N = 235	18 (7.7)	217 (92.3)		4 (1.7)	109 (46.4)	143 (65.9)	14 (6.0)
Total anomalous pulmonary venous connection, N = 6	0 (0)	6 (100)	10.0	1 (16.7)	6 (100)	4 (66.7)	1 (16.7)
Anomalies pulmonary artery, N = 229	18 (7.9)	211 (92.1)	NA	3 (1.3)	103 (45.0)	139 (65.9)	13 (5.7)
Atrial septal defects, N = 1265	97 (7.7)	1168 (92.3)		17 (1.3)	420 (33.2)	510 (43.1)	65 (5.2)
Atrial septal defects, N = 1265	97 (7.7)	1168 (92.3)	12.9		420 (33.2)	510 (43.1)	65 (5.2)
Ventricular septal defects, N = 393	28 (7.1)	365 (92.9)		0 (0)	59 (15.0)	98 (26.6)	10 (2.6)
Ventricular septal defects, N = 393	28 (7.1)	365 (92.9)	12.9–68.6	0 (0)	59 (15.0)	98 (26.6)	10 (2.6)

Data not shown for rare fetal cardiac anomaly classifications in our cohort: heterotaxy, complex defects, and atrioventricular septal defects (n = 11).

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RVOTO, left ventricular outflow tract obstruction; *NA*, not available; *NICU*, neonatal intensive care unit; *RVOTO*, right ventricular outflow tract obstruction.

^aData extrapolated from previous studies ^{7,8};

^bColumn denominator is number of patients with NICU admission or 5-min Apgar score documentation available;

^cN corresponds to number of neonates with respective category of anomaly.

Composite neonatal outcome by completed weeks of gestation for attempted vaginal delivery and planned cesarean delivery in cases of fetal cardiac anomalies^a

TABLE 2

Fetal cardiac anomaly classification	Completed gestation, wk						P value
	34–36 6/7 n/N _t ^b (%)	37–37 6/7 n/N _t ^b (%)	38–38 6/7 n/N _t ^b (%)	39–39 6/7 n/N _t ^b (%)	40–40 6/7 n/N _t ^b (%)	41 n/N _t ^b (%)	
Conotruncal defects, N = 107 ^c	N ₁ = 17	N ₂ = 18	N ₃ = 23	N ₄ = 30	N ₅ = 12	N ₆ = 7	
Attempted vaginal delivery							
Spontaneous labor, N = 47	5/6 (83.3)	2/6 (33.3)	6/12 (50.0)	3/11 (27.3)	4/8 (50.0)	0/4 (0)	.04 ^d
Labor induction, N = 42	7/9 (77.8)	5/6 (83.3)	3/6 (50.0)	6/14 (42.9)	1/4 (25.0)	2/3 (66.7)	.07
Planned CD, N = 18	2/2 (100)	4/6 (66.7)	3/5 (60.0)	3/5 (60.0)	–	–	–
P value ^e	1.00	.35	1.00	.49	–	–	
LVOTO, N = 83	N ₁ = 15	N ₂ = 17	N ₃ = 24	N ₄ = 19	N ₅ = 7	N ₆ = 3	
Attempted vaginal delivery							
Spontaneous labor, N = 34	5/8 (55.6)	3/5 (60.0)	5/8 (62.5)	3/7 (42.9)	2/4 (50.0)	2/2 (100)	1.00
Labor induction, N = 29	0/2 (0)	3/4 (75.0)	7/10 (70.0)	5/10 (50.0)	2/2 (100)	1/1 (100)	.24
Planned CD, N = 20	4/5 (80.0)	4/8 (50.0)	1/4 (25.0)	1/2 (50.0)	1/1 (100)	–	–
P value	.27	.83	.44	1.00	.62	–	
RVOTO, N = 72	N ₁ = 15	N ₂ = 9	N ₃ = 15	N ₄ = 17	N ₅ = 10	N ₆ = 6	
Attempted vaginal delivery							
Spontaneous labor, N = 28	2/3 (66.7)	3/5 (60.0)	1/4 (25.0)	1/4 (25.0)	3/8 (37.5)	0/4 (0)	.07
Labor induction, N = 27	4/7 (57.1)	3/3 (100)	3/7 (42.9)	2/6 (33.3)	1/2 (50.0)	1/2 (50.0)	.38
Planned CD, N = 17	3/5 (60.0)	0/1 (0)	4/4 (100)	1/7 (14.3)	–	–	–
P value	1.00	.29	.16	.79	–	–	
Congenital anomalies pulmonary artery, N = 235	N ₁ = 95	N ₂ = 41	N ₃ = 31	N ₄ = 46	N ₅ = 10	N ₆ = 12	
Attempted vaginal delivery							
Spontaneous labor, N = 105	33/46 (71.7)	7/13 (53.9)	4/13 (30.8)	10/20 (50.0)	3/6 (50.0)	2/7 (28.6)	<.01 ^d
Labor induction, N = 60	17/22 (77.3)	5/15 (33.3)	2/6 (33.3)	3/12 (25.0)	2/2 (100)	0/3 (0)	<.01 ^d
Planned CD, N = 70	17/27 (63.0)	9/13 (69.2)	3/12 (25.0)	7/14 (50.0)	1/2 (50.0)	1/2 (50.0)	.19
P value	.54	.19	1.00	.36	.71	.71	

Fetal cardiac anomaly classification	Completed gestation, wk					P value
	34–36 6/7	37–37 6/7	38–38 6/7	39–39 6/7	40–40 6/7	41
Atrial septal defects, N = 1265	n/N _t ^b (%)	n/N _t ^b (%)	n/N _t ^b (%)	n/N _t ^b (%)	n/N _t ^b (%)	n/N _t ^b (%)
Attempted vaginal delivery	N ₁ = 243	N ₂ = 167	N ₃ = 256	N ₄ = 316	N ₅ = 189	N ₆ = 94
Spontaneous labor, N = 589	86/125 (68.8)	29/70 (41.4)	27/100 (27.0)	41/155 (26.5)	19/96 (19.8)	6/43 (14.0)
Labor induction, N = 371	29/49 (59.2)	17/42 (40.5)	24/61 (39.3)	34/110 (30.9)	13/65 (20.0)	11/44 (25.0)
Planned CD, N = 305	52/69 (75.4)	28/55 (50.9)	23/95 (24.2)	12/51 (23.5)	8/28 (28.6)	1/7 (14.3)
P value	.18	.52	.12	.58	.56	.44
Ventricular septal defects, N = 393	N ₁ = 65	N ₂ = 43	N ₃ = 76	N ₄ = 119	N ₅ = 64	N ₆ = 26
Attempted vaginal delivery						
Spontaneous labor, N = 206	21/41 (51.2)	1/17 (5.9)	7/38 (18.4)	6/57 (10.5)	3/39 (7.7)	2/14 (14.3)
Labor induction, N = 106	2/12 (16.7)	2/14 (14.3)	1/13 (7.7)	5/36 (13.9)	2/21 (9.5)	0 (0)
Planned CD, N = 81	5/12 (41.7)	5/12 (41.7)	4/25 (16.0)	3/26 (11.5)	0/4 (0)	0 (0)
P value	.10	.06	0.77	0.93	1.00	.57

Data not shown for rare fetal cardiac anomaly classifications in our cohort: heterotaxy, complex defects, and atrioventricular septal defects (n = 11).

CD, cesarean delivery; LVOTO, left ventricular outflow tract obstruction; RVOTO, right ventricular outflow tract obstruction.

^aComposite neonatal outcome (serious respiratory morbidity, sepsis, birth trauma, hypoxic ischemic encephalopathy/asphyxia/seizures, neonatal mortality);

^bn/N_t corresponds to number of composite neonatal outcome (n) over number of neonates (N_t) for respective gestational age group and labor intervention category—percentage of this fraction is presented in parenthesis—N₁–6 corresponds to total number of neonates by gestational age group with each category of fetal cardiac anomaly;

^cN corresponds to number of individual neonates with fetal cardiac anomaly classification further divided into labor intervention;

^dStatistically significant;

^eRow P value Fisher exact test for comparison between spontaneous labor, labor induction, and planned CD—column P value Cochran-Armitage trend test for comparison across weeks of gestation.