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Delivery room emergencies in critical congenital heart diseases

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Abstract

Introduction: Transition of fetus to postnatal is a complex process. Fetal congenital heart disease is one of the commonest anomaly which is encountered presently in clinical setups.

Objective: To determine the mode of delivery in pregnancies complicated by complex fetal congenital heart disease (CHD).

Methods: three-year retrospective cohort study at different hospitals (2017 to 2020). Cases of complex fetal CHD (n=126) were compared with non-anomalous singleton infants ⩾500 g to measure ratio of emergency intrapartum caesarean section (CS), vaginal delivery and preterm delivery.

Results: Fetal CHD did not increase emergency C-section rates in nulliparous women, in multiparous pregnancies C-section was significantly increased. Intrapartum C-section is significantly greater in fetal CHD than non-anomalous controls.

Conclusion: Emergency C-section is higher in fetal CHD, attributed to a higher rate of C-section for non-reassuring fetal status and seen mostly in multiparous women.

Keywords: CHD, Fetal, Heart, Diseases.

Introduction

Fetal to postnatal life transformation is a complicated procedure. About 4-10% of new-borns need some form of aid even congenital heart disease is not present (**1**). Fetal congenital heart disease (CHD) is a common structural abnormality which is experienced commonly in clinical practice. Prenatal assessment involving fetal and maternal health in general and detailed information on fetal heart structure, function and hemodynamic in certain are critical for planning the delivery. Many researchers argue that, elective preterm delivery does not show any benefit if sustained fetal arrhythmia is absent (**2**). It is oftenly contended that normal delivery should be the aim in fetal CHD, conserving C-section for obstetric complications, with which we would agree. Past studies have shown that high rates of successful vaginal delivery can be achieved in complex fetal CHD without hampering neonatal outcomes(**3**).

Advances in fetal echocardiography using high-resolution ultrasound and serial imaging have conducted an increased number of fetuses diagnosed with congenital heart disease. Clinical course in utero and at delivery can now be forecasted, and as a result, fetal medicine experts are being asked to think the fetus as a patient and the transition to postnatal life an important part of care (**4**). Although many studies show that fetal diagnosis includes outcomes for patients with critical congenital heart disease by preventing postnatal instability, certain populations of patients with congenital heart disease continue to have high mortality rates due to compromise that begins in the delivery room(**5**). New-borns with hypoplastic left heart syndrome (HLHS) or d-transposition of the great arteries (d-TGA) with restrictive foramen ovale or intact atrial

septum, Ebstein's anomaly, tetralogy of Fallot with absent pulmonary valve or arrhythmias with hydrops have all been reported to have poor outcomes if diagnosed prenatally(**6**). For these fetuses, subspecialty care must begin in the delivery room to affect morbidity and improve survival. Some free-standing children's hospitals have included delivery suites to house high-risk deliveries, but expectant women may have increased risk if deliveries are performed at facilities that rarely provide this service(**8**). For fetuses with little or no risk for requiring specialized delivery room care, delivery at the local hospital is recommended (**9**).

Irrespective of this argument, past researches on this issue have focused on outcomes in prenatally diagnosed vs postnatally diagnosed CHD.The outcome of pregnancies complicated by severe fetal CHD compared with healthy non-anomalous pregnancies has been poorly studied(**10**). Given the high rates of associated major anomalies in CHD and the scope for clinician anxiety in such complicated anomalies, it seems reasonable to ask whether cases of CHD fare similarly in labor and delivery to healthy controls (**11**). We proposed a retrospective cohort study over a 2-year study period at a busy maternal–fetal medicine department, with an increased number of prenatally diagnosed fetal CHD. Our aim was to examine the mode of delivery in cases of fetal CHD vs non-anomalous controls, particularly the rate of emergency intrapartum CS (**12**).

Methods

We conducted a retrospective cohort study of pregnancies with known fetal CHD delivering in a single tertiary obstetric centre between 15 February2018 and 15 February 2020. Data was collected from maternal-fetal medicine department and a level III neonatal intensive care unit of different hospitals i.e. Services Hospital, Mumtaz Bakhtawar Hospital and Kot Khawaja Saeed Hospital. The decision for CS is made by a consultant obstetrician (specialist with at least 10 years clinical experience) in every case. The overall rate of caesarean delivery in those hospitals during the 3-year study period was 20%.

In the hospital all major fetal cardiac defects were managed by a multidisciplinary team including a maternal–fetal medicine specialist, neonatologist, paediatric cardiologist and nurse specialist. Basically normal delivery is the aim, with elective CS conserved for obstetric complications or fetal arrhythmias. Induction of labor is considered from 39 weeks on individual basis, depending on the cardiac lesion, parity, Bishop’s score and patient distance from the hospital. Babies born with major CHD are stabilized in the neonatal intensive care unit and a neonatal echocardiogram is performed as soon as possible by a paediatric cardiologist. Following initial management, infants are transferred to paediatric cardiology department for definitive intervention.

Cases of fetal cardiac disease were identified from an institutional fetal anomaly register, with obstetric outcomes extracted from computerized hospital records. Indications for delivery for all CS are recorded prospectively in this database. All major fetal congenital heart defects in singleton infant’s $\geqslant$500 g were eligible. Isolated ventricular septal defects and premature atrial contractions, which would not be anticipated to influence prognosis, were excluded.

Among the fetal cardiac cases, we also examined the relationship between mode of delivery and Apgar scores, cord pH and early neonatal death ($\leqslant$7 days). This was a retrospective analysis of anonymised hospital data; as such, this study was deemed exempt from IRB approval.

Statistical analysis was performed using Spss version 2.0. For the primary case–control analysis, odds ratios (ORs) and 95% confidence intervals (CIs) were calculated.

Median values for Apgar scores and cord pH demonstrated evidence of non-normality on Shapiro–Wilk's test. Other categorical data were compared using Fisher's exact test. As such, these data were compared using the Mann–Whitney test for non-parametric data. Two-tailed P-values were used throughout and the level of significance was considered 5%.

Results

126 cases of major fetal CHD were delivered between 2017 and 2020. The major heart defects were hypoplastic left heart syndrome (19% of total CHD), Fallot's/double-outlet right ventricle (12%), atrioventricular septal defect (16%) and cardiomyopathy (10%) and transposition (9%), with smaller numbers of other major fetal CHD. Of the 126 CHD cases, three miscarriages were before fetal viability and there were nine antepartum stillbirths at ⩾24 weeks, yielding 114 (91%) surviving cases of major fetal CHD at ⩾24 weeks, which were observed in the current analysis. The total rate of major congenital anomaly associated with fetal CHD was 32% (37/114). In one-third of cases, the associated anomaly was trisomy 21, with heterotaxy syndrome (19%) and congenital diaphragmatic hernia (11%) the next most commonly associated major abnormalities.

observing mode of delivery in the 114 cases of major CHD reaching viability, the overall number of emergency intrapartum C-section was significantly large in the fetal CHD cases as compare to the non-anomalous controls (21% vs 13.5%, OR 1.7, P=0.035). We found no difference in the number of emergency intrapartum caesarean section for other clinical complications (OR 1.47, P=0.40).

On examining by parity, we found that in nulliparous women there was no difference in the overall rate of intrapartum CS or in the rate of CS for NRFHT in fetal CHD cases compared with controls. However, in multiparous women, major fetal CHD is predominantly increased both total intrapartum CS (OR 2.4, P=0.018) and CS performed for NRFHT (OR 5.4, P=0.003. The intrapartum CS ratio for multiparous controls with and without previous CS were 25% and 6.8%, respectively. In multiparous CHD cases, the ratio of intrapartum CS in women with an unscarred uterus was 17%.

Data was also analysed adjusting for cases complexed by a coexisting major congenital anomaly (37/114). The number of major non-cardiac malformations in laboring women with fetal CHD was 30%, with no difference in women that ultimately delivered vaginally vs those who needed CS (P=0.61). When intrapartum CS ratios were adjusted for the presence of major non-cardiac abnormalities, the number of emergency CS in isolated fetal CHD was 21% (16/78), which is higher than the extent in non-anomalous controls, although it did not reach statistical significance (OR 1.7, 95% CI: 0.7 to 3.9; P=0.06).

Consequently a sub analysis was performed on the cases of major fetal CHD who labored. Fetal CHD cases delivered through normal delivery were not associated with poorer Apgar scores or cord pH levels when measured with those delivered by CS; indeed, both the 1- and 5-min Apgar scores were statistically higher in the normal delivery group, although such a small difference is not clinically appreciated. There were two intrapartum stillbirths in this group, both in the normal delivery group and both with major complex cardiac defects. Thus, the overall live-birth rate among laboring women was 96% (102/114). Analysis of early neonatal survival demonstrated a 5.2% early neonatal mortality (8/109), with no difference between the vaginal and CS cases (P=0.58). For cases of isolated major fetal CHD, the rate of early neonatal death was 1% (1/67).

Discussion

In this large case–control study, singleton pregnancies complicated by complex fetal CHD have a significantly higher rate of emergency intrapartum C-section compared with non-anomalous controls. For both total fetal CHD cases and for cases of isolated CHD only, the intrapartum C-section ratio was 21%, which represents a 50% increase in risk over the control population (13.5%). Furthermore, this increased rate was due almost a substantially higher number of C-section performed for non-reassuring fetal status; no increase in intrapartum C-section for other indications in the CHD cases was demonstrated. As such, this increased rate cannot be ascribed to a higher rate of failed induction, a possible explanation given that induction of labor was more prevalent in the CHD cases.

Analysis by parity found that emergency C-section in fetal CHD was 19 to 23% across all parity groups. Against the higher background rate in nulliparous women, this represents no significant increase. However, emergency C-section ratio were significantly higher in multiparous women in the presence of fetal CHD; indeed, the rate of C-section for NRFHT was increased more than fivefold. This significant difference remained even when women with a previous C-section were excluded. Thus, the difference does not appear to be related to the effect of previous C-section in laboring multiparous women. To our knowledge, these findings have not previously been reported.

Several theories may explain the higher rate of emergency C-section in cases of complex fetal CHD. One possible culprit is the higher rate of preterm delivery, with CHD cases more than three times as likely to deliver at <37 weeks in the present study(**13**). Even in healthy fetuses, preterm delivery is associated with higher rates of CS, although evidence regarding any protective benefit remains conflicting. Additionally, obstetricians' perceptions and anxiety may contribute to a higher CS rate in these high-risk cases. In the Californian study, cases of fetal CHD had higher C-section rates when the diagnosis was known compared with undiagnosed cases. Although this suggests that a known diagnosis of fetal cardiac disease may influence clinician practice, there was also a higher rate of non-cardiac defects in the prenatally diagnosed subgroup. The recent large study from New York did not substantiate this hypothesis; rates of intrapartum C-section were not higher in the prenatally diagnosed cases (16.5%) vs the undiagnosed cases (15%), although rates of prelabor CS were ⩾30% in both groups.

Few previous authors have examined obstetric outcomes in pregnancies complicated by major fetal cardiac disease. In 2008, an Australian study described 92 fetuses with mixed-severity cardiac disease. Although the authors reported no maternal morbidity, specific data for gestation and mode of delivery were not reported (**14**). Subsequently, a Californian study analysed 329 surviving neonates with CHD; in more than half of cases (55%), diagnosis was not made prenatally. Overall C-section rates for diagnosed and undiagnosed fetal CHD in this study were 66% and 50%, respectively. Unfortunately, outcomes for healthy controls were not reported in this study; thus, it is not possible to determine whether the rate of C-section in undiagnosed CHD cases is higher than the background rate, although 50% certainly appears high. Very recently, Landis et al. reported outcomes for 993 neonates with CHD, of which 68% were diagnosed prenatally. The overall C-section rate in this study was 45%, of which approximately one-quarter were performed for NRFH (**15**).

The current study also supports the high rates of live birth in fetuses with CHD reported by other authors. Only 12% of major fetal CHD in our study were delivered by elective C-section and, among laboring women, we did

not demonstrate poorer Apgar scores or arterial blood gases in women delivered vaginally. While the higher rate of early neonatal death in the vaginal subgroup is notable (6% vs 0%), when perinatal mortality is adjusted for the presence of major non-cardiac defects, we see no difference in the mode of delivery.

Finally, it remains possible that fetuses with complex CHD have a lower inherent tolerance for labor and consequent higher intrapartum CS rates. A huge Swedish population-based Swedish study of >6300 fetuses with CHD reported significantly higher rates of meconium aspiration, fetal distress and CS when compared with the general population. Additionally, a Japanese study reported higher rates of both abnormal fetal heart rate patterns and intrapartum C-section for fetal indications in 116 CHD cases. Although we agree that vaginal delivery offers advantages, particularly for the mother, the present study, which found a higher rate of intrapartum CS for NRFHT in cases of major CHD, appears to lend further credence to the hypothesis that major fetal CHD is associated with a higher rate of emergency CS (**16**).

We acknowledge several limitations with the present study. The retrospective nature of this study introduces the possibility of bias. Second, this was a study in a single obstetric centre, which practices a standard protocol of labor management; our total C-section rate (20%) and rate of prelabor C-section for fetal CHD (12%) were low by international standards. Although we believe this draws the C-section rates in CHD cases into sharper focus, it is likely that such pronounced differences would not be demonstrated in units with high baseline caesarean rates. Third, although we have attempted to discriminate between isolated fetal CHD and cases with extra cardiac anomalies, this distinction is limited to anomalies identified antenatal or immediately postnatal. Last, while we have provided data on early neonatal outcomes for laboring women with fetal CHD, our study is underpowered to examine the effect of mode of delivery on perinatal outcomes.

Nonetheless, this is a large 3-year study that, unlike most previously published work, compares outcomes in fetal CHD with non-anomalous controls. It is reassuring to find no difference in Apgar scores or cord pH in normal delivered CHD cases compared with those requiring emergency C-section, underscoring the safety of vaginal delivery for these women. The finding that intrapartum C-section rates are higher in cases of major fetal CHD should assist clinicians in counselling women faced with this complex situation.

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