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Research paper

Outcomes of implementing cardiac risk stratification and perinatal care recommendations for prenatally diagnosed congenital heart disease

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Abstract

Introduction: Prenatal diagnosis of congenital heart disease (CHD) can best achieve decreases in perinatal mortality and improvements in preoperative clinical status when combined with risk stratification and active perinatal management. This study assessed implementation of a four-tier risk classification system for prenatally diagnosed CHD based on the need for emergent neonatal cardiac intervention (ENCI), each with specific perinatal care recommendations.

Material and methods: Prenatally diagnosed fetuses with CHD were risk stratified, and fetal, perinatal and postnatal data were collected with the primary aim to assess risk stratification accuracy. Secondary aims assessed adherence to perinatal management recommendations and evaluation of outcomes compared to a non-risk stratified historical cohort.

Results: The study cohort comprised 84 patients prenatally diagnosed with CHD with assigned ENCI level. This included ENCI level 1 ($n = 4$), level 2 ($n = 16$), level 3 ($n = 50$), and level 4 ($n = 14$). Cardiac intervention was performed emergently in 8 cases (9.5%), non-emergently in neonates in 56 cases (67%), and before one year of age in an additional 15 cases (18%). Our classification system correctly risk stratified 90.4% (76/84) of infants with CHD based on their need for neonatal cardiac intervention. No ENCI level 1 or 2 risk stratified infants required neonatal intervention, and only one of the 50 ENCI level 3 cases required an unanticipated ENCI. Thus, the ENCI classification system correctly identified 98.5% (69/70) of patients who did not require emergent neonatal cardiac intervention in the first 48 hours of life. For perinatal management recommendations, the delivery notifications were appropriately sent 94% of the time, neonatology was present for 99% of recommended deliveries, and PGE was started according to recommendations in 93% (53/57) of ENCI level 3 and 4 cases. The ENCI cohort (2012-2014) compared to an earlier non-risk stratified cohort of 146 patients (2008-2011) showed no difference in survival ($p = 0.66$) or hospital length of stay ($p = 0.19$).

Conclusions: Implementation of our prenatal risk stratification system accurately predicted the need for emergent neonatal cardiac intervention, and adherence to perinatal management recommendations was high.

Key words: risk stratification, prenatal diagnosis, congenital heart disease, fetal echocardiography, cardiac intervention.

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Introduction

A prenatal diagnosis of congenital heart disease (CHD) has been shown to decrease perinatal morbidity and improve clinical status prior to cardiac surgery [1-7]. However, it is not just the diagnosis itself, but the perinatal management that is required to improve outcomes in the fetus with CHD. Particularly in cases of severe CHD, which require cardiac intervention within the first year of life (https://www.cdc.gov/ncbddd/heartdefects/cchd-facts.html). The highest risk cases are those forms of CHD that require emergent surgical and/or catheter-based cardiac intervention in the first 48 hours of life. Identification and risk stratification of these fetuses is important, as the postnatal presentation can vary dramatically based on CHD type. Examples include d-transposition of the great arteries (dTGA) with adequate atrial communication that is stable with good oxygen saturations to that of dTGA with restrictive atrial septum (RAS) that require emergent catheter balloon atrial septostomy (BAS). Similarly, total anomalous pulmonary venous return (TAPVR) without obstruction can be stable with no immediate hemodynamic collapse, or the pulmonary venous return can be obstructed and require emergent cardiac surgery. Proactive and individualized perinatal planning allows for a coordinated delivery and immediate postnatal care according to risk stratification.

Prenatal risk stratification in CHD is based on fetal echocardiography findings for which several systems are currently in use and/or are being actively studied [8-12]. At our own institution a method of risk stratification was developed and implemented for prenatally diagnosed CHD to stratify their risk for requiring emergent neonatal cardiac intervention (ENCI), incorporating a four-tier classification system and coordinated perinatal care plan (Table 1) [2]. ENCI level 1 comprises patients with mild CHD who do not require neonatal cardiac evaluation or intervention. ENCI level 2 patients require neonatal cardiac evaluation but not neonatal cardiac intervention. Level 3 patients require neonatal cardiac intervention and are typically prostaglandin dependent; thus, they require a higher level of NICU acuity to provide immediate postnatal care. ENCI level 4 probably require immediate postnatal intervention, either catheter based or surgical, within $\leq$ 48 hours of life. The ENCI guidelines have specific care recommendations depending on the anticipated level of perinatal risk regarding initiation of prostaglandin E1 (PGE), mode of delivery (MOD), neonatal intensive care unit (ICU) acuity for delivery, neonatology presence at delivery, and cardiac team notification. There are other important considerations such as maternal co-morbidities, fetal extracardiac anomalies, genetic condition, and gestational age, but here we focused on the CHD diagnosis as the primary basis for risk stratification.

We implemented this system at our institution in 2011 and after several years of refining the classifications and care plans we set out to determine: 1) accuracy of risk stratification to predict ENCI level 1-4, 2) adherence to perinatal management recommendations, and 3) outcomes compared to an earlier non-risk stratified cohort.

Material and methods

A retrospective study was performed for patients prenatally diagnosed with CHD, who underwent ENCI classification, and who were cared for at a large Fetal Maternal Center (FMC) established and managed by Children's Hospital Los Angeles

Table 1. Emergent Neonatal Cardiac Intervention (ENCI) classification system

Class	Description	PGE	Mode of delivery an issue	NICU acuity level	Neonatology present in delivery room	Cardiology, CT Surgery, CTICU, OR/cath lab on standby	Examples
ENCI 1	Low risk	No	No	Low	No	No	ASD, VSD, mild PS
ENCI 2	Medium risk	No	No	Mid	Possibly	No	CAVC, TOF/PS, truncus arteriosus
ENCI 3	Medium-high risk	Likely	Possibly	High	Yes	Possibly	HLHS, dTGA/VSD, PA/IVS
ENCI 4	High risk	Likely	Yes	High	Yes	Yes	dTGA/RAS, obstructed TAPVR

ASD – atrial septal defect, CAVC – complete atrioventricular canal defect, CT – cardiothoracic, CTICU – cardiothoracic intensive care unit, dTGA – dextro-transposition of the great arteries, HLHS – hypoplastic left heart syndrome, NICU – neonatal intensive cardiac unit, OR – operating room, PGE – prostaglandin E, PA/IVS – pulmonary atresia with intact ventricular septum, PS – pulmonary stenosis, RAS – restrictive atrial septum, TAPVR – total anomalous pulmonary venous return, TOF – tetralogy of Fallot, VSD – ventricular septal defect

(CHLA) and University of Southern California (USC) Obstetrics. We used a consecutive sampling of our FMC RedCap database over a three-year period to enroll pregnant women with a prenatal diagnosis of CHD with assigned ENCI classification of 1 to 4 based on previously published criteria (Table 1) [8, 13]. All patients had a complete fetal echocardiogram evaluation at the FMC or CHLA fetal cardiology clinic [9]. If a patient had multiple fetal echocardiograms and/or ENCI levels, then the one closest to delivery was used for analysis. While patients were delivered at a number of different affiliated obstetrical centers, only patients seen at CHLA for postnatal neonatal evaluation and/or cardiac intervention were included in the study. All patients included in the study had their diagnoses confirmed by postnatal echocardiography. Patients were excluded for gestational age < 36 weeks, insufficient prenatal diagnosis and/or delivery plan data, or lack of ENCI classification. This study was approved by the Institutional Review Boards of USC and CHLA.

The primary aim of the study was to evaluate the ENCI cohort for accuracy of risk stratification to the appropriate ENCI level based on the neonatal outcomes. The clinical outcomes were obtained by review of delivery room, neonatal ICU and cardiac ICU records to confirm which patients required cardiac evaluation or intervention and the timing of this. The prenatal ENCI designation was only deemed accurate if the evaluation/intervention occurred within the time period specified by each level.

A secondary aim of the study looked at adherence to the ENCI system care plans by comparing delivery and perinatal care records to the recommendations. Neonatal cardiac intervention was defined as cardiac surgery or catheterization within 30 days of birth. PGE therapy was considered appropriate when started after birth when recommended in the fetal consultation note. MOD was confirmed by review of obstetric delivery records as vaginal delivery with or without induction and C-section (CS) planned or emergent. NICU acuity was based on the delivery centers associated with our children's hospital. Delivery notification was based on the fetal-maternal center notification email. Specific communication to cardiology, cardiothoracic surgery, catheterization lab, and CTICU teams was counted only if documented in the chart.

Another secondary aim was to compare outcomes for the ENCI cohort compared to an earlier cohort of patients with prenatally diagnosed CHD admitted to CHLA for heart surgery from 2008-2011, who were not risk stratified (pre-ENCI). In the pre-ENCI cohort, individual recommendations were made for patients on a case-by-case basis. While neonatology was present for all high-risk CHD deliveries, there was no formal process to notify surgical or catheterization teams postnatally or have them on standby. The two cohorts were evaluated for differences in timing of intervention, hospital length of stay, 30-day survival, and survival to discharge home. Groupwise differences of normally distributed continuous variables were evaluated by Student's T-test and non-normally distributed variables by Wilcoxon Rank Sum. The difference in 30-day survival between the pre-ENCI and ENCI eras was tested using Fisher's Exact test. A p value of < 0.05 was considered significant.

Results

The institutional database included 189 unique fetal cases of whom 98 had CHD from 2014-2016, with the others having non-cardiac anomalies or other reasons to be followed by FMC. Of those, 84 were assigned an ENCI classification (86%) and included in the study. See Table 2 for details of all cardiac diagnoses for each ENCI category. The study cohort consisted of 62% male infants, and patients were born at an average gestational age of 37.8 ±1.8 weeks. The three most common diagnoses were tetralogy of Fallot (TOF) (n = 23, 27%), ventricular septal defect (VSD) (n = 22, 26%), and hypoplastic left heart syndrome (HLHS) (n = 16, 19%). Genetic disorders were present in 21% of cases (n = 18) including heterotaxy syndrome (n = 6) and chromosomal anomalies (n = 9), with the most common being 22q11deletion, 'DiGeorge syndrome' (n = 6). Extracardiac malformations were found in 22% (n = 19) of cases including renal anomalies (n = 6), situs anomalies (n = 5), and central nervous system anomalies (n = 4). Of note, there were 5 patients with antibody-mediated congenital complete heart block (CCHB) assigned ENCI levels 3 and 4.

Neonatal cardiac intervention was performed in 67% of patients (56/84), of whom 53 had neonatal surgery at a mean age of 8.7 days and 15 had neonatal cardiac catheterization at a mean age of 6.9 days (12 of whom also had surgery). Figure 1 gives details of the cardiac interventions performed for each ENCI category.

Table 2. Cardiac diagnoses for each ENCI level in the study cohort

ENCI level 1 (n = 4)	ENCI level 2 (n = 16)	ECNI level 3 (n = 50)	ENCI level 4 (n = 14)
VSD (2) ASD (1) Possible CoA (1)	TOF (8) CoA + VSD (5) VSD + PS (1) CoA + VSD + AS (1) CAVC (1)	HLHS (10) SV + PA or PS (6) PA/IVS (5) TOF (2) TOF + PA (2) TOF + PA + MAPCAs (2) TGA + VSD + PS (2) TGA + SV (2) EA (2) CHB (2) Tricuspid atresia + VSD + PS (1) Truncus Arteriosus (1) Shone's syndrome + CoA (1) CCTGA + SV + AS (1) IAA + VSD (1) DORV + TGA (1) SV + TAPVR (1) CAVC + PA (1) Critical PS (1)	DTGA (5) HLHS + RAS (3) CHB (2) SV + PA + TAPVR (1) SV + IAA + AS + TAPVR (1) SV + PA + AS (1) 2nd Degree AVB + LQTS (1)

AS – aortic stenosis, ASD – atrial septal defect, AVB – atrioventricular block, CAVC – complete atrioventricular canal defect, CCTGA – congenitally corrected transposition, CHB – complete heart block, CoA – coarctation, DORV – double outlet right ventricle, dTGA – dextro-transposition of the great arteries, EA – Ebstein's anomaly, HLHS – hypoplastic left heart syndrome, IAA – interrupted aortic arch, MAPCAs – major aortopulmonary collaterals, PA/IVS – pulmonary atresia with intact ventricular septum, PS – pulmonary stenosis, RAS – restrictive atrial septum, SV – single ventricle, TAPVR – total anomalous pulmonary venous return, TOF – tetralogy of Fallot, VSD – ventricular septal defect

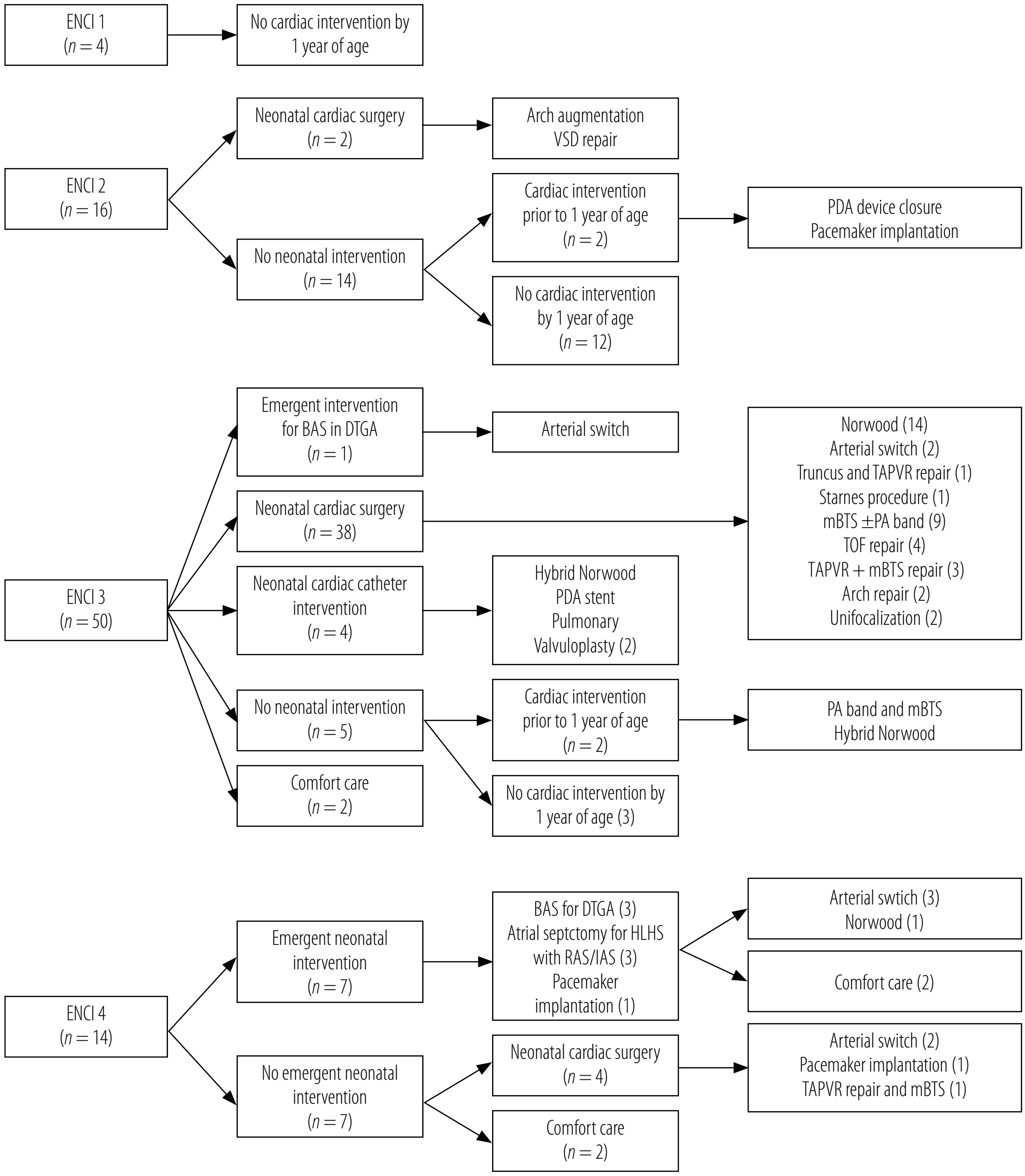

Figure 1. Flow chart of cardiac interventions performed for each ENCI category

AS – aortic stenosis, ASD – atrial septal defect, AVB – atrioventricular block, CAVC – complete atrioventricular canal defect, CCTGA – congenitally corrected transposition, CHB – complete heart block, CoA – coarctation, DORV – double outlet right ventricle, dTGA – dextro-transposition of the great arteries, EA – Ebstein's anomaly, HLHS – hypoplastic left heart syndrome, IAA – interrupted aortic arch, MAPCAs – major aortopulmonary collaterals, PA/IVS – pulmonary atresia with intact ventricular septum, PS – pulmonary stenosis, RAS – restrictive atrial septum, SV – single ventricle, TAPVR – total anomalous pulmonary venous return, TOF – tetralogy of Fallot, VSD – ventricular septal defect

The ENCI classification system correctly identified 98.5% (69/70) of patients who did not require neonatal cardiac intervention (Table 3). No ENCI level 1 or 2 patients needed emergent intervention. ENCI classification correctly identified 90% (7/8) of those who did require ENCI in the first 48 hours of life. One ENCI level 3 case required unanticipated emergent neonatal cardiac intervention. This case involved a patient with dTGA and VSD who developed a RAS postnatally requiring BAS. The 14 patients classified as ENCI level 4 are detailed in Table 4. Seven patients required emergent neonatal cardiac intervention including 3 of 5 dTGA patients with RAS requiring BAS, three patients with HLHS and RAS requiring surgical atrial septectomy, and one patient with congenital complete heart block requiring emergent pacemaker placement.

Postnatal adherence to ENCI care recommendations are detailed in Table 3. Practice variability across the recommended ENCI care guidelines was low. Delivery notifications were appropriately sent 94% of the time, and the neonatology team was present for 99% of recommended deliveries. PGE was started according to recommendations in 87% of cases (67/77), including 93% (53/57) of ENCI level 3 and 4 cases. In fact, PGE was initiated in all cases in which it was specifically recommended

Table 3. Adherence to Emergent Neonatal Cardiac Intervention (ENCI) Guidelines

Class	Description	Number of cases	ENCI	Appropriate PGE therapy	Delivery notification sent	Elective C/S or elective induction	GA ≥ 38 weeks at delivery, if C/S or induction	Neonatal resuscitation team at delivery
ENCI 1	Low risk	4	0	3/4 (75%)	3 (75%)	n/a	n/a	n/a
ENCI 2	Medium risk	16	0	11/16 (69%)	16 (100%)	n/a	n/a	16 (100%)
ENCI 3	Medium-high risk	50	1 (2%)	42/46 (91%)	46 (92%)	23/50 (46%)	30/31 (97%)	49 (98%)
ENCI 4	High risk	14	7 (50%)	11/11 (100%)	14 (100%)	5/14 (36%)	13/14 (93%)	14 (100%)
Total		84	8/84(9.5%)	67/77 (87%)	79/84 (94%)	28/64 (44%)	43/45 (96%)	79/80 (99%)

C/S – caesarean section, GA – gestational age, PGE – prostaglandin E

Table 4. ENCI level 4 clinical outcomes

Diagnosis	Met ENCI Level 4 Criteria	Required ENCI	Emergent Cardiac Intervention < 48 hours	Neonatal cardiac intervention > 48 hours and ≤ 30 days	Clinical outcome
dTGA	Yes	Yes	BAS	ASO	Discharge home
dTGA	Yes	Yes	BAS	ASO	Discharge home
dTGA	Yes	Yes	BAS	ASO	Discharge home
dTGA	Yes	No		ASO	Discharge home
dTGA	Yes	No		ASO	Discharge home
dTGA*with VSD	No	Yes	BAS	ASO	Discharge home
HLHS w/RAS	Yes	Yes	Atrial septectomy	Norwood-Sano	Discharge home
HLHS w/IAS	Yes	Yes	Atrial septectomy	None	Multi-organ failure, comfort care
HLHS w/IAS	Yes	Yes	Atrial septectomy	None	Multi-organ failure and CVA, comfort care
CHB	Yes	Yes	PM implant		Discharge home
CHB	Yes	No		PM implant	Discharge home
2nd degree AV block, LQTS	Yes	No		None	Discharge home
TAPVR-mixed, single ventricle, pulmonary atresia, heterotaxy	Yes	No		TAPVR repair and AP shunt	Post op death due to multi-organ failure
Dextrocardia, TAPVR-obstructed, CAVC, AS, IAA	Yes	No		None	No intervention, comfort care
Dextrocardia, CAVC, primitive single ventricle with poor function, single outflow was BAV with AS	Yes	No		None	No intervention, comfort care

AP – aortopulmonary, AS – aortic stenosis, ASO – arterial switch operation, BAS – balloon atrial septostomy, BAV – bicuspid aortic valve, CAVC – complete atrioventricular canal defect, CHB – complete heart block, CVA – cerebral vascular accident, dTGA – dextro-transposition of the great arteries, HLHS – hypoplastic left heart syndrome, IAA – interrupted aortic arch, IAS – intact atrial septum, LQTS – long QT syndrome, PGE – prostaglandin E, PA/IVS – pulmonary atresia with intact ventricular septum, PM – pacemaker, RAS – restrictive atrial septum, TAPVR – total anomalous pulmonary venous return, VSD – ventricular septal defect.
**This patient was categorized as ENCI level 3, but required ENCI.*

based on prenatal fetal echocardiography findings. PGE was started after birth without prenatal recommendation in 30% (6/20) of ENCI level 1 and 2 cases. It is not known why PGE was started in 4 of the 6 cases, because it was not indicated by the clinical status of the patient. However, the other two cases in which PGE was started for clinical suspicion of arch obstruction were ultimately diagnosed with coarctation, making the administration of PGE appropriate in these cases. Evaluation for mode of delivery showed that 23 of 50 patients had an elective C-section (CS) or induction as recommended. However, only 5 of the 14 ENCI level 4 cases (36%) were delivered by recommended scheduled CS. This was due to spontaneous labor prior to their scheduled CS date in 5 cases (36%) and patient preference for induction of vaginal delivery in 4 cases (28%).

An earlier cohort of patients with CHD was identified that included 146 patients admitted to CHLA between 2008 and

Table 5. Comparison of non-risk-stratified to ENCI risk-stratified cohort

Parameter	Pre-ENCI cohort non-risk stratified ($n = 146$)	ENCI cohort ($n = 84$)	*P* value
Mean birth weight (SD)	2958 ±620 g	2870 ±561 g	0.28
Extra-cardiac malformations	32 (20%)	19 (22%)	0.9
Chromosomal anomaly or genetic syndrome	32 (22%)	18 (21%)	0.93
Median age at surgery (IQR)	5 days (3-9)	5 days (3-8)	0.37
Median length of hospital stay (IQR)*	18 days (10-45)	23 days (11-50)	0.19
Survival to 30 days*	89.9%	91.8%	0.66
Survival to discharge*	85.4%	84.9%	0.93

*IQR – interquartile range, SD – standard deviation. *These analyses were based on the ENCI Cohort that underwent cardiac intervention or died prior to intended cardiac intervention (n = 78).*

2011 for cardiac intervention in the first year of life. The earlier cohort of patients was CHD was not risk stratified using the ENCI classification (Pre-ENCI). Patients who received comfort care due to severe co-morbidities were excluded from survival and length of stay analysis, but patients who died prior to cardiac intervention with intention to treat were included. When compared to an earlier cohort of patients who were not risk stratified (pre ENCI), there was no difference seen in age at surgery, 30-day survival, hospital length of stay, or survival to discharge home. See Table 5 for a complete comparison of the two cohorts.

Discussion

Risk stratification and active perinatal management for cases of prenatally diagnosed CHD are now being employed with the goal of improving perinatal mortality and preoperative clinical status. We implemented the ENCI classification system at our institution and were able to correctly risk stratify 90.4% (76/84) of infants with CHD based on their need for neonatal cardiac intervention. No ENCI level 1 or 2 risk-stratified infants required emergent intervention, and only one of the 50 ENCI level 3 cases required an unanticipated emergent neonatal cardiac intervention. The ENCI classification system correctly identified 98.5% (69/70) of patients who did not require emergent neonatal cardiac intervention in the first 48 hours of life. The only case in which unanticipated emergent neonatal cardiac intervention was required involved a patient with dTGA and VSD, who underwent BAS within 48 hours for a RAS.

In the level 4 risk-stratified group only 50% (7/14) of neonates required emergent neonatal intervention (Table 4). Of the other 7 patients, 2 were provided comfort care only. Therefore, 5 patients were assigned an ENCI level 4 classification that did not ultimately have an intervention in the first 48 hours: 2 DTGA, 1 CHB, one second-degree AV block, and one complex single ventricle with TAPVR. The ability to accurately predict risk need for emergent postnatal BAS in dTGA with RAS physiology based on prenatal fetal echocardiography imaging alone has been well studied, but it remains a challenge for risk stratification and has resulted in many stratification systems automatically categorizing dTGA at the highest risk level [10, 14-16]. This is because the fetal echocardiographic features of the atrial septum, foramen ovale, pulmonary venous Doppler, and ductus arteriosus used to predict RAS have low sensitivities and specificities [17]. Until fetal echocardiography markers for predicting RAS in DTGA are more predictive of neonatal compromise, we anticipate a degree of over-assignment (or levelling up) to the ENCI level 4 category in order to prevent potential serious perinatal complications and preoperative morbidities [8, 9, 12].

PGE administration after birth was started in 30% of lower ENCI level cases without prenatal recommendation. We suspect that newborns with known CHD are more likely to be treated reactively with PGE regardless of recommendations, which may be similar to the previously reported higher rates of preoperative intubation in prenatally diagnosed CHD patients [6]. Our current care guidelines do not recommend intubation for ENCI levels 1-3 if not clinically indicated. However, we do recommend intubation often for ENCI level 4 cases due to the increased risk for rapid decompensation prior to anticipated intervention. It is our hope that reviewing these guidelines regularly with the neonatal and obstetrical staff at delivery hospitals caring for newborns with severe CHD will ensure that PGE administration and intubation are only performed in the appropriate situations and not reactively without purpose.

Adherence to mode of delivery was difficult to determine due to the wide range of indications for undergoing induction of labor versus Cesarean section. ENCI level 3 patients for whom MOD was not anticipated to be a major variable were frequently delivered by C/S or induction (44%). Multiple studies have shown that rates of both emergency C/S and labor induction are higher in CHD patients and that prenatal diagnosis of CHD is associated with preterm delivery [13, 18]. There has been a concerted effort in the obstetric community to avoid unnecessary C/S and decrease preterm delivery (< 37 weeks) [16, 19, 20]. This trend was supported by our findings that the ENCI level 3 and 4 patients had a gestational age at delivery that was on average > 38 weeks.

While this study showed no difference in outcomes compared to an earlier cohort of patients with CHD, who were not risk stratified, it did lead directly to several improvements in our perinatal management strategies. First, we standardized our communications regarding pregnancies complicated by prenatal diagnosis of CHD by creating a formalized document used among various specialty teams for pending deliveries.

Second, we increased our compliance with ENCI level assignments and documentation in fetal consultation reports. Third, transport teams were coordinated ahead of time for the highest risk deliveries to facilitate immediate transfer of the newborn to the children's hospital. Lastly, treatment was streamlined by obtaining transport and procedural consents during prenatal visits, prior to the anticipated delivery dates.

Our study has several important limitations, including the small sample size and fixed datapoints in the medical record, which may have decreased our ability to show true differences in clinical outcomes (Type II error). We could not fully assess adherence to the recommendation for delivery in a hospital with an appropriate level of NICU acuity because our cohort was predominantly delivered at the delivery hospital (with a level III NICU) affiliated with our children's hospital. This may make our data less applicable to other healthcare systems with a wider range of NICU levels. We also could not adequately assess the effectiveness of delivery notifications to cardiology, CT surgery, or the ICU team because we did not document these communications in official medical records at the time of our study. Lastly, while comfort care was not a formal part of the ENCI system, it was discussed because it related to patient care and may warrant a separate classification category.

Conclusions

In conclusion, the ENCI risk stratification system was successfully implemented with a high accuracy of care assignment, and adherence to perinatal care recommendations was generally high. While early implementation of risk stratification did not show a difference in clinical outcomes, this study led to immediate improvements in our care management strategies, including standardized communications and documentation, prenatal coordination of transport teams, and prenatal consenting for transport and anticipated emergent postnatal procedures.

Conflict of interest

The authors declare no conflict of interest.

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