RESEARCH PAPER

# Outcomes of Infants with Prenatally Diagnosed Congenital Heart Disease Delivered in a Tertiary-care Pediatric Cardiac Facility

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*Received: August 29, 2014; Initial review: October 21, 2014; Accepted: July 15, 2015.*

**Objective:** To report short-term outcomes of infants with prenatally diagnosed Congenital Heart Disease (CHD) delivered in a tertiary-care cardiac facility.

**Design:** Retrospective study.

**Setting:** Tertiary-care referral hospital.

**Participants:** Children with prenatally diagnosed CHDs who underwent delivery at study centre during the period January 2008 - December 2013 were included. Outcomes tracked from hospital records and direct follow-up.

**Results:** Of the 552 fetuses diagnosed to have CHD, 121 (22%) were delivered at the study centre. Fetuses undergoing a planned delivery were diagnosed in late gestation (mean gestational age 31.5 $\pm$ 5.1 wk). 74 fetuses (61.2%) had simple CHD and rest were complex. 96 (79.3%) neonates received cardiac care; 30 (24.8%) required surgery while 5 received catheter-based interventions. 11 patients underwent surgery on follow-up. Neonatal survival in cardiac care group was 93.8%; on follow-up (12.5 $\pm$ 13.1 mo); 83 (86.4%) of these infants were alive. All infants undergoing neonatal surgery or catheter-based interventions survived. 25 patients (20.6%) received comfort care (Complex CHD, associated co-morbidities); 14 (56%) survived neonatal period and 6 (24%) were alive on follow-up.

**Conclusions:** Infants with prenatal diagnosis of CHD and planned delivery in a cardiac facility had satisfactory immediate outcomes, expecially in those receiving specialized post-natal cardiac care.

**Keywords:** *Congenital heart disease, Fetal echocardiograpgy Planned delivery, Prenatal diagnosis*.

Congenital Heart Diseases (CHD) occur in 2.5 to 3 per 1000 live births and is a major cause of morbidity and mortality in newborn babies [1,2]. About 7% of all neonatal deaths are attributable to congenital malformations of which about 25% are due to severe forms of CHD [3-5]. Differences exist in the rates of prenatal diagnosis of CHD across various populations and this can have an impact on the live birth prevalence of complex CHD [6-12]. Recent studies report favorable impact of prenatal diagnosis on neonatal outcomes for infants with critical CHD [13-15]. Practice guidelines and training programs for obstetric sonographers on screening fetal heart have significantly improved prenatal detection of CHD [16,19].

There is limited data regarding the impact of prenatal diagnosis on outcomes of CHD in developing countries [20]. Recently, selected centres have started performing neonatal heart surgery with outcomes comparable to the developed world [21]. In the setting of significant logistic hurdles for transport and delivering prompt neonatal cardiac care, prenatal diagnosis may enable a more efficient utilization of the limited health care resources for the postnatal treatment of critical CHD [4]. This study reports the short-term outcomes of infants with prenatally diagnosed CHD undergoing a planned delivery in a tertiary-care pediatric cardiac facility in Kerala, India.

*Accompanying Editorial: Pages 845-46.*

## Methods

The study was conducted in a tertiary-care pediatric cardiology centre in Kochi after Institutional Ethical clearance. This study was a retrospective review of the short-term outcome of all infants with prenatally diagnosed CHD delivered in the study centre during the period January 2008 to December 2013. The data was collected in a database and outcomes were tracked by direct follow-up and review of hospital records. The other outcomes of all prenatally diagnosed CHD (pregnancies not culminating in live births, delivered elsewhere and not evaluated, lost to follow-up and normalization of findings

on post-natal echocardiography were recorded in the database, but not included in the analysis.

Variables included in the analysis were maternal age, gestational age during fetal echocardiography indication of referral, maternal systemic illness and fetal risk factors. CHD was classified as simple (those amenable for anatomic repair without need for right ventricle to pulmonary artery conduit with low probability of re-operations) and complex (those which require palliative procedures in the lines of univentricular heart or anatomic repair with conduit with high possibility of re-operations).

All expectant families were counseled regarding the prognosis of CHD, management options and the expected outcomes based on current literature and institutional experience. In India, the legal limit for termination of pregnancy is 20 weeks gestation [22]. The option of planned delivery in a cardiac centre was discussed whenever appropriate. The final decision regarding the management was taken by the families in consultation with the referring obstetrician.

Immediate outcomes were categorized on the basis of the type of care provided after birth – cardiac or comfort care groups. In the cardiac care group, details of medical management and definitive therapeutic management (surgery or trans-catheter intervention) were noted. In the comfort care group, reasons for the decision for comfort care (complexity of CHD or presence of associated anomalies) were noted. Immediate neonatal outcomes (survived/died) were recorded for both groups. For survivors, details of follow-up were recorded including procedures, complications and deaths.

*Statistical analysis*: Statistical analysis was done using SPSS. Fischer's exact test was used for comparison of continuous variables. *P* value <0.05 was considered as significant.

## RESULTS

A total of 2087 women were referred for fetal echocardiography during the study period; 552 (26.4%) were diagnosed to have fetal CHD. Of these,121 (22%) were delivered in our centre and constitute the study group. The other outcomes included pregnancies not culminating in live-births (44%), delivered elsewhere and not evaluated (22%), lost to follow-up (5%) and normalization of findings on postnatal echocardiography (7%). The mean (SD) maternal age was 27.1 (5.1) years. Fetuses delivering in the study center were diagnosed to have CHD at a later stage of gestation; mean (SD) gestational age 31.5 (5.1) weeks. The indications for referral included suspicion of CHD on screening (81.8%), maternal illness (12.4%), extra-cardiac anomalies (4.1%) and family history of CHD (1.7%). Maternal risk factors were reported in 26%, most common being diabetes mellitus (17.4%). Thirteen fetuses (10.7%) were conceived through artifical reproductive techniques.

Forty-eight (39.7%) fetuses were delivered vaginally; the rest were delivered by cesarian section (53.7%) or instrumental delivery (6.6%). Sixty-three (52.1%) were males. Nineteen fetuses (15.7%) were delivered before term. The mean (SD) birth weight was 2.7 (0.6) kg; 33.1% were small for gestational age, 6.6% large and the rest were approriate for gestational age.

***Table* I** summarizes the CHDs diagnosed by fetal echocardiography. There was a statistically signifcant association between the type of CHD and delivery in cardiac facility (Simple 28.5% *vs* Complex 17.7%; *P*=0.004). There was an increasing trend towards planned delivery during the study period (***Fig.* 1**). Post-natal echocardiography was concordant with the fetal diagnosis in 107 patients (88.4%); minor variations not affecting outcomes were noted in 7 (5.8%) infants. Major discordance from fetal diagnosis was noted in 7 patients (5.8%) (***Table* II**).

Ninety-six (79.3%) of the neonates received cardiac care while the remainder were included in the comfort care group. Thirty infants ( 24.8%) needed surgery in the neonatal period, while 11 (9%) underwent surgery on followup. ***Table* III** summarizes the details of surgical procedures performed. Five (4.1%) infants needed

**TABLE I** CONGENITAL HEART DEFECTS DIAGNOSED BY FETAL ECHOCARDIOGRAPHY (*N* = 121)

| *Cardiac Diagnosis* | *Number (%)* |
|---|---|
| Single Ventricle | 18 (14.9) |
| Double Outlet Right Ventricle | 18 (14.9) |
| Tetralogy of Fallot | 17 (14) |
| Tricuspid valve anomalies | 11 ( 9.1) |
| Transposition of great arteries | 8 (6.6) |
| Coarctation of aorta | 6 (5) |
| Pulmonary valve stenosis | 5 (4.1) |
| Left-to-right shunts | 5 (4.1) |
| Hypoplastic left heart syndrome | 5 (4.1) |
| Tachy-arrhythmias with no CHD | 4 (3.3) |
| Pulmonary atresia with intact septum | 3 ( 2.5) |
| Truncus Arteriosus | 3 (2.5) |
| Ventricular disproportion | 3 (2.5) |
| Brady-arrhythmias with no CHD | 2(1.7) |
| Cardiac Tumors | 2 (1.7) |
| Miscellaneous | 11 ( 9.1) |

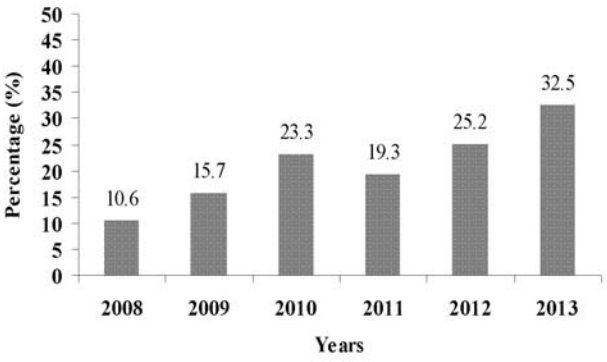


**FIG. 1** *Trends in planned delivery in the study center 2008-13.*

catheter-based interventions, 3 in the neonatal period and 2 on follow-up.

Patients who were offered comfort care had either a very complex CHD (68%) or had significant associated co-morbities (48%) or both (32%). Co-morbidities included associated genetic syndromes (6 patients) and extra-cardiac anomalies (6 patients).

There were a total of 17 deaths (14%) in the neonatal period; 6 in the cardiac group and 11 in the comfort care group. All the deaths in the cardiac care group occurred in patients who were managed medically. None of the infants who needed neonatal surgery or catheter interventions died. Of the 90 survivors in the cardiac care group, 83 were alive on follow-up (12.5 ± 13.1 months); 5 died and 2 were lost to follow-up. Of the 5 deaths, 2 occurred in operated patients while the other 3 had significant extra-cardiac anomalies. ***Fig.* 2** summarizes the immediate and follow-up outcomes of the study patients.

## DISCUSSION

This study reports the outcomes of 121 infants with prenatally diagnosed CHD undergoing planned delivery in a cardiac facility. The majority of neonates (79.3%) thus delivered received post natal cardiac care tailored according to the clinical indication. The immediate outcome of infants who underwent surgery or catheter-based interventions was excellent with no neonatal mortality.

Several studies from developed countries have reported the overall favorable impact of pre-natal diagnosis on outcomes for critical CHDs [13-15]. Few studies have studied the impact of site of delivery of these

**TABLE II** MAJOR DISCORDANCE IN FINDINGS BETWEEN PRENATAL AND POSTNATAL ECHOCARDIOGRAPHY

| *Prenatal Diagnosis* | *Postnatal diagnosis* |
|---|---|
| Tricuspid regurgitation, RVH | Critcal Pulmonic stenosis with intact septum |
| Tetralogy of Fallot | Truncus arteriosus Type 1 |
| Truncus Arteriosus | Double outlet RV; Transposed great vessels, VSD |
| Tetralogy of Fallot | Transposition of great arteries with VSD |
| Tetralogy of Fallot | Transposition of great arteries with VSD and Coarctation |
| Double outlet RV with transposed great vessels | Corrected transposition of great arteries, VSD, Pulmonary atresia |
| Double aortic arch | Left aortic arch with PDA |

*RVH – Right ventricular hypertrophy; VSD – Ventricular septal defect; PDA – patent ductus arteriosus.*

**TABLE III** LIST OF SURGICAL PROCEDURES PERFORMED IN THE CARDIAC CARE GROUP

| *Newborn period ( n=30)* | | *Follow-up (n = 11)* | |
|---|---|---|---|
| Arterial Switch Operation | 11 | Bidirectional Glenn shunt | 3 |
| Aorto-pulmonary shunt | 9 | Pulmonary artery band | 2 |
| Coarctation of aorta repair | 2 | Intra-cardiac repair | 2 |
| Arterial switch with arch repair | 2 | Truncus repair | 1 |
| VSD closure with arch repair | 2 | Intra-cardiac repair with conduit | 1 |
| Stage 1 Norwood Operation | 2 | AV canal defect repair | 1 |
| TAPVC repair | 1 | VSD closure | 1 |
| Permanent pacemaker implantation | 1 | | |

*VSD – Ventricular Septal Defect; TAPVC – Total anomalous pulmonary venous connection.*

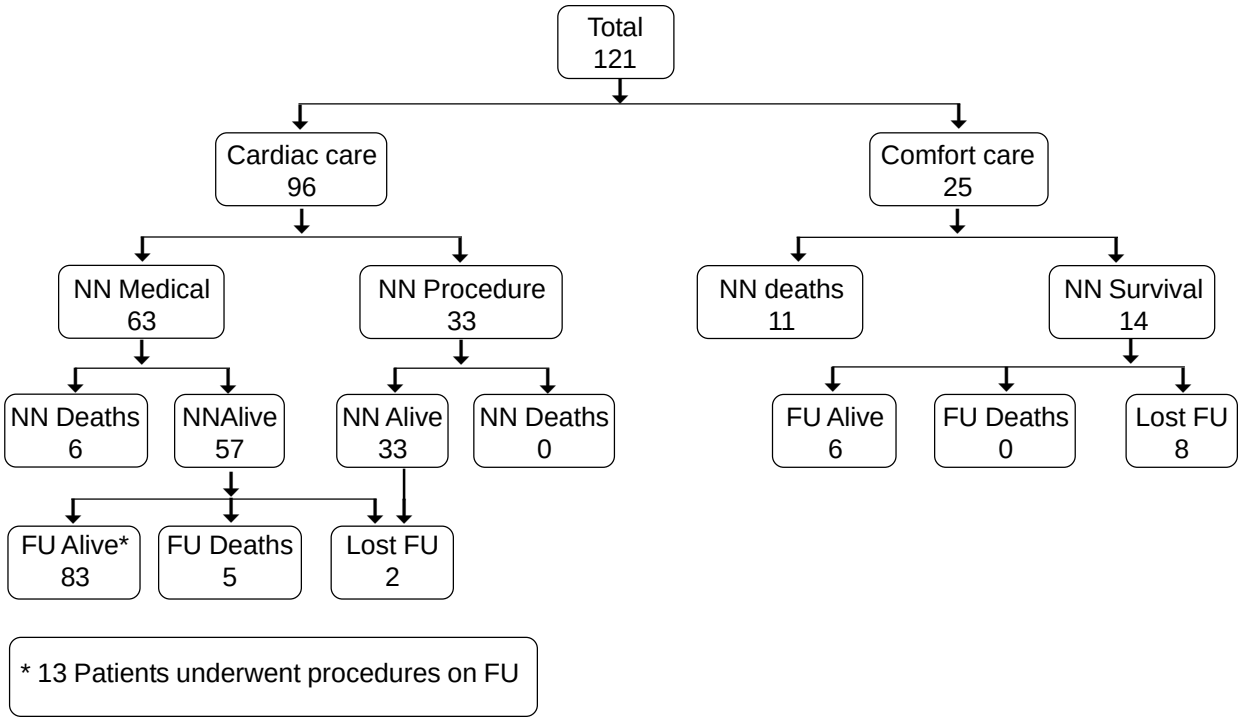


**FIG. 2** *Outcomes of prenatally diagnosed CHD delivered in the study center* (*N*= 121) *NN – Neonatal*; *FU – Follow-up.*

infants on the outcomes. A recent study from United Kingdom had reported the feasibility of delivering infants with prenatally diagnosed CHD outside specialist cardiac centers [23]. A significant proportion of neonates in this study were transferred to a cardiac facility within the first few days of life (median 2 days). Neonatal survival reported in this study was 90%. However, the health care systems in developing countries are vastly different. Difficulties in getting a prompt pediatric cardiology consult and echocardiography, delay in initiation of life saving medications like prostaglandin infusion, and logistic hurdles involved in the transport to a cardiac facility can signifcantly compromise the fragile hemodyanamic status of a neonate with critical CHD. This can adversely impact the pre-operative status of these infants and thereby the outcomes after intervention. The strategy of planned delivery in a cardiac facility as reported in this study potentially overcomes all these logistic difficulties and can result in an improved pre-operative clinical state, thus favorably impacting outcomes. Planned delivery also enables the baby and mother to be admitted in the same facility, which is a signifcant pyschological advantage for families.

The outcomes in the comfort care group was poor. This highlights the need to diagnose such complex CHDs in an earlier stage of pregnancy. An earlier diagnosis (<20 weeks) would have given more options to the expectant family including consideration for termination of pregnancy [22]. This highlights the need for training of obstetric sonographers on fetal heart screening including the four-chamber view and outflow tracts.

The study is limited by its retrospective design, and the possibility that the study population may have been affected by referral bias. The study has not compared the outcomes of prenatally diagnosed CHD with those who were diagnosed postnatally. However, the practical advantages of such a approach are apparent from the data, even without a comparative analysis.

In conclusion, prenatal diagnosis of CHD and planned delivery in a cardiac facility results in excellent immediate outcomes in neonates receiving specialized post-natal cardiac care. There is a pressing need to diagnose complex CHD in early pregnancy through better ultrasound screening, providing more options to expectant families.

*Contributors*: BV: designed the study, edited and approved the manuscript and shall act as guarantor for the manuscript; TDC: collected and analyzed data and prepared the manuscript; AJ, RR: collected and analyzed data; AS analyzed and interpreted the data; RK: critically reviewed and approved the final draft of the mansucript.
*Funding*: None; *Competing interests*: None stated.

**WHAT IS ALREADY KNOWN?**

- Pre-natal diagnosis of CHD is feasible with high diagnostic accuracy.

**WHAT THIS STUDY ADDS?**

- Prenatal diagnosis of CHD and planned delivery in a cardiac facility results in excellent immediate outcomes in neonates receiving specialized post-natal cardiac care.

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