Evidence-Based Fetal Referrals for Congenital Heart Surgery

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Abstract

Background. The outcomes of fetal referrals to congenital heart disease centers for delivery and postnatal surgery prior to discharge over a two-year period were reviewed. Cost differences between fetal referrals and neonatal transports were investigated.

Methods. A retrospective chart review was conducted on 17 fetal referrals to two congenital heart disease centers from 01/01/2007 to 12/31/2008. The two centers were contacted to obtain their neonatal transport charges.

Results. Of the 17 fetal referrals, 10 patients underwent congenital heart surgery prior to postnatal discharge. Only one patient who underwent surgery died. Third party payers saved approximately $13,600 or $36,600 in neonatal transport costs to these centers.

Conclusions. There was only one death of a patient with hypoplastic left heart syndrome and a restrictive atrial septum, which has a poor prognosis. There was a significant cost differential between fetal referral and neonatal transport.

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Introduction

Congenital heart disease occurs in 9 of 1000 livebirths.[1] Critical congenital heart disease, which requires interventional catheterization or surgery in the first year of life, occurs in about 25% of these children. Antenatal diagnosis of critical congenital heart disease and referral of the mother for confirmation and planned delivery at a congenital heart disease center in these cases may allow for optimum outcomes[2,3] with referrals in favor of the best centers.[4]

A prenatal diagnosis of congenital heart disease did not result in savings in cost, length of hospitalization, or survival in the relatively small geographic area of New England, which lends itself to rapid patient transfer.[5] However, a study of three referral centers in Northern California reported that most infants without a prenatal diagnosis of congenital heart disease require one or more transports to congenital heart disease centers with ventilation and prostaglandin therapy.[6]

Allen and colleagues[7] reported superior outcomes in patients requiring congenital heart surgery by selectively referring to high-volume surgical centers based on published or "apparent" low mortality rates for specific congenital cardiac surgical procedures. Congenital heart surgery has not been done at our center since 1998. All of our patients are referred to major congenital heart surgery centers. Using Allen and colleagues' model for superior surgical outcomes, our institution developed and used an evidence-based referral system for in utero diagnosis of congenital heart disease likely to require early intervention.

Parents are counseled about the intervention and the option of referral to an evidence-based demonstrated congenital heart surgical center in cases most likely to require intervention prior to postnatal discharge. These centers are long distance, 200 miles (center 1) and 900 miles

(center 2), from our institution. Patients that we believe will have a good outcome are referred to center 1. Center 2 is nationally known as one of a few centers with the lowest mortality (less than 10%) for complex congenital heart disease requiring the Norwood procedure. Therefore, patients likely to require the Norwood procedure are referred to center 2.

This study was a two-year retrospective review of fetal referrals undergoing post-natal congenital cardiac surgery prior to discharge. Mortality outcomes and the cost differences between fetal referral and neonatal transport to the two centers were investigated.

Methods

From 01/01/2007 to 12/31/2008, a perinatologist referred 81 patients to one of two pediatric cardiologists for a comprehensive fetal echocardiogram. The pediatric cardiologists obtained the fetal echocardiograms at the perinatologist's office with one of two certified obstetrical sonographers. The fetal echocardiograms were performed on a Voluson Expert 730 with an AC 2-5 transducer until May, 2008, when a GE-E8 with a 4C transducer was used.

A retrospective chart review of these 81 patients was completed. Seventeen patients were identified whose parents chose evidence-based referral to one of these two centers. The following data were extracted from these 17 charts: date of comprehensive fetal echocardiogram, fetal gestational age, maternal age, pediatric cardiologist, reason for referral, chromosomal anomaly, extracardiac anomaly, echocardiography findings, congenital heart surgical center where the patient was referred, and outcome.

The two centers were contacted to obtain their charges for neonatal transport. The cost of driving to these centers was estimated. The average cost of a motel at center 2 was estimated using an American Automobile Association (AAA) travel book. The cost of flying to center 2 was obtained from an AAA travel agent.

Results

Ten patients underwent congenital heart surgery procedures prior to discharge. Two patients underwent other interventions. Two patients did not require intervention prior to discharge. One patient's mother had premature rupture of membranes and delivered at her local hospital. Two patients were lost to follow-up.

The two patients who were lost to follow-up were seen initially at center 1. One had aortic valve stenosis, a dilated left ventricle, and a patent foramen ovale. The other one had severe tricuspid valve hypoplasia, pulmonary valve stenosis, infundibular stenosis, a restrictive ventricular septal defect, a secundum atrial septal defect, and a hypoplastic right ventricle. There was no record of them being admitted to that center after birth.

One patient with an absent pulmonary valve, severe pulmonary insufficiency, right atrial enlargement, and right ventricular hypertrophy was seen at center 1. That patient has not had any intervention. One patient with a small left ventricle was thought to have a probable severe coarctation on fetal echocardiogram. Only a secundum atrial septal defect was found after birth at center 2, and that patient did not require intervention.

The fetus, whose mother had premature rupture of membranes with delivery at the local hospital, had a complete atrio-ventricular block secondary to maternal lupus. The infant was followed by a pediatric electrophysiologist at center 1 and has not had a pacemaker implanted yet.

One patient with pulmonary valve atresia underwent radio-frequency perfor-

ation and balloon dilation prior to discharge from center 1. That patient had undergone two subsequent balloon dilations. One patient with levo-transposition of the great arteries, a ventricular septal defect, pulmonary valve stenosis, and complete atrioventricular block underwent permanent pacemaker implantation prior to discharge from center 1.

Details of the 10 patients who underwent congenital heart surgery prior to discharge are shown in Table 1. In some cases, subsequent congenital heart surgical procedures were known. The patient with hypoplastic left heart syndrome and a restrictive atrial septum, which has a poor prognosis, was profoundly cyanotic at birth. The patient was immediately evaluated and taken to the operating room for a hybrid procedure. This was the only patient who died. Thus, a 10% mortality was observed for our 10 patients who underwent congenital heart surgical procedures.

In review of the expected costs of travel for the parents, there were no flights from our city to center 1. The cost of driving to either center was estimated using 15 miles per gallon at $3 per gallon. The parents should not have had to stay overnight when going to center 1. The parents would incur transportation costs at the initial evaluation and again when returning to center 1 for delivery. However, the second trip was likely not a cost difference as at least one parent would have to drive to center 1 after delivery at our institution if the baby underwent a neonatal air transport to center 1. The estimated cost for both parents to drive to center 1 was $110.

Similarly, at least one parent would have to drive or fly to center 2 after delivery at our institution if the baby underwent a neonatal air transport to center 2. As mentioned, the cost of a motel at center 2, and the cost of roundtrip airfare from our city to the closest airport to center 2 was per AAA. There was an additional estimated cost of a roundtrip shuttle from the airport to center 2. The estimated costs for both parents to drive to center 2, stay at a motel for four nights, and eat for five days was $1,210. The estimated costs for both parents to fly to center 2, use an airport shuttle, stay at a motel for two nights, and eat for three days was $1,447.80. Center 1 charged approximately $13,600 for neonatal air transport. Center 2 charged approximately $36,600 for neonatal air transport. Table 2 shows comparative costs.

Discussion

In our retrospective review to determine the effects of evidence-based fetal referral on mortality and cost, a reduction in expected mortality and lower total cost was observed with the use of evidence-based referral for fetally detected congenital heart disease requiring surgery prior to discharge.

In a retrospective review, Yeager et al.[8] reported that cardiac patients transported from adjacent obstetric facilities compared to cardiac patients transported from other inpatient medical facilities were more likely to have been diagnosed prenatally with more complex disease and had higher mortality. A study from Boston Children's Hospital found that a prenatal diagnosis of hypoplastic left heart syndrome or transposition of the great arteries improves the preoperative condition of these patients, but it may not significantly improve preoperative mortality or early postoperative outcome among neonates managed at a tertiary center.[9] However, a University of California-San Francisco study of patients with hypoplastic left heart syndrome found that all patients diagnosed prenatally and who underwent surgery survived.[10] Of 38 patients diagnosed postnatally who under-

Table 1. Congenital heart surgical procedures and outcomes.

Center	Gestational Age (weeks/days)	Diagnosis	Initial Intervention	Subsequent Interventions	Outcome
1	24	Tricuspid valve atresia, VSD[1], Normal great arteries	Blalock-Taussig shunt	Bidirectional Glenn shunt	Alive
1	27	VSD[1], DORV[2], d-malposition[3], Pulmonary valve stenosis	Blalock-Taussig shunt	Blalock-Taussig shunt, Bidirectional Glenn shunt	Alive
2	35	Coarctation	Repair with patch	None	Alive
2	23	HLHS[4]	Norwood	None	Alive
2	34	Hypoplastic aortic arch, Coarctation, Small aortic valve	Norwood	Coarctation repair (elsewhere)	Alive
2	23	Tricuspid valve atresia, VSD[1], Normal great arteries	Blalock-Taussig shunt, Atrial septectomy	Hemi-Fontan, Fenestrated Fontan	Alive
2	36	HLHS[4]	Norwood	Hemi-Fontan, Fenestrated Fontan	Alive
2	25	HLHS[4], Restrictive atrial septum	Atrial stent (embolized), Stent removal, Atrial septectomy, Bilateral branch pulmonary artery bands	None	Dead
2	23	DILV[5], Right atrioventricular valve atresia, l-TGA[6]	Atrial septectomy, Pulmonary artery band	Hemi-Fontan, Fenestrated Fontan	Alive
2	28	TV atresia, VSD[1], d-TGA[7], Hypoplastic aortic arch	Damus-Kaye-Stansel procedure	Bidirectional Glenn shunt	Alive

[1]VSD (ventricular septal defect), [2]DORV (double-outlet right ventricle), [3]d-malposition (dextro-malposition of the great arteries), [4]HLHS (hypoplastic left heart syndrome), [5]DILV (double-inlet left ventricle), [6]l-TGA (levo-transposition of the great arteries), [7]d-TGA (dextro-transposition of the great arteries).

Table 2. Cost comparisons.

Center 1		
Fetal Referral		**Neonatal Transport**
Gasoline	$80	$13,600
Food	$15/person	
Center 2		
Fetal Referral		**Neonatal Transport**
Gasoline or Airline Airport Shuttle	$360 $438.90/person $50/person	$36,600
Motel	$100/night	
Food	$45/person/day	

went surgery, only 25 survived. Patients diagnosed prenatally had a lower incidence of preoperative acidosis, tricuspid regurgitation, and ventricular dysfunction. They were less likely to need preoperative inotropic medications or bicarbonate.

Sharland et al.[11] in a retrospective review, found difficulties in diagnosing coarctation prenatally. Of 87 fetuses, coarctation was diagnosed correctly in 54, suspected but unproved in 24, and overlooked prenatally in 9. They concluded that although a combination of echocardiographic features can identify aortic arch anomalies in the fetus, none, either alone or in combination, could distinguish between real and false positive cases, particularly in late gestation. As reported above, one false positive case was observed in our study.

Jenkins et al.[12] developed a consensus-based method of risk adjustment for in-hospital mortality among children younger than 18 years after surgery for congenital heart disease (designated RACHS-1). Their data sources were the Pediatric Cardiac Care Consortium (PCCC) and hospital discharge data sets. The PCCC includes 32 congenital heart surgery centers. The three statewide hospital discharge data sets were from Illinois, Massachusetts, and California.

Coarctation repair less than 30 days of age is a risk category 2 procedure with an expected mortality rate by PCCC data of 3.8% and hospital discharge data of 3.3%. The Blalock-Taussig shunt and the pulmonary artery band are risk category 3 procedures with an expected mortality rate of 8.5% and 6.8% respectively. Atrial septectomy, a risk category 4 procedure, has an expected mortality rate of 19.4% and 16.4% respectively. Finally, the Norwood operation and the Damus-Kaye-Stansel procedure are risk category 6 procedures. The mortality rate in this category for PCCC data is 47.7% and for hospital discharge data is 41.5%.

In our study, one patient was in risk category 2, three patients in risk category 3, two patients in risk category 4, and four patients in risk category 6. Using the data of expected mortality in RACHS-1, a mortality of 22-26% was expected. Only one death was observed in a patient with hypoplastic left heart syndrome and a restrictive atrial septum undergoing an emergent hybrid procedure for a mortality of 10%.

As mentioned previously, Copel et al.[5] did not find that a prenatal diagnosis of congenital heart disease resulted in cost savings. Although Friedberg et al.[6] stated that the need to transport critically ill

neonates to a referral center potentially compromises their hemodynamic stability and is costly, they did not report the actual costs of transports to the three Northern California centers or the transport distances.

Our study was limited by being retrospective. Cases may have been omitted or missed, but we are unaware of any such cases. Lack of follow-up data in two cases in such a small series was a limitation. Our institution is small with only two pediatric cardiologists making for possible limitations in the wider use of this referral pattern. Statistical power was limited by not having large numbers of fetuses with critical congenital heart disease which require surgery prior to discharge. However, only one patient died of those with follow-up whose parents chose to deliver at the evidence-based congenital heart surgery centers recommended by the pediatric cardiologist.

In conclusion, lower mortality than expected was demonstrated in this small group of patients. By selectively referring patients requiring the highest risk procedures to a center with the lowest mortality, our patients achieved mortality rates as good as the best centers and better than the RACHS-1 estimates. Centers in the RACHS-1 study probably do not refer to other centers.

Third party payers would incur the costs of a perinatology consult, a pediatric cardiology consult, a fetal echocardiogram, and possibly a congenital heart surgeon's consult at the time of the initial referral. However, these costs would be much less for third party payers than those for neonatal transport. The parents incurred additional travel expense by having to travel to the congenital heart disease center for confirmatory fetal echocardiogram and arranging delivery at the referral center. The costs for the parents were greater if they traveled to the more distant center.

Potentially with these types of data, third party payers would consider paying the travel expenses for the parents, which would be less expensive and a more cost effective use of resources. Based on the perceived advantage in outcomes and demonstrated cost savings, our institution continues to refer patients fetally diagnosed with congenital heart disease and likely to require early intervention for delivery at the evidence-based surgical centers.

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