Survivors of Congenital Diaphragmatic Hernia Repair Face Barriers to Long-Term Follow-Up Care

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Short Title: **Barriers to Outpatient Care following Congenital Diaphragmatic Hernia Repair**

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Abstract

Background

Congenital diaphragmatic hernia (CDH) carries high morbidity and mortality, and survivors commonly have neurodevelopmental, gastrointestinal, and pulmonary sequela requiring multidisciplinary care well beyond repair. We predict that following hospitalization for repair, CDH survivors face many barriers to receiving future medical care.

Methods

A retrospective review was conducted of all living CDH patients between ages 0 to 12 years who underwent repair at Riley Hospital for Children (RHC) from 2010 through 2019. Follow-up status with specialty providers was reviewed, and all eligible families were contacted to complete a survey regarding various aspects of their child's care, including functional status, quality of life, and barriers to care. Bivariate analysis was applied to patient data ($p<0.05$ was significant) and survey responses were analyzed qualitatively.

Results

After exclusions, 70 survivors were contacted. Thirty-three (47%) were deemed lost to follow up to specialist providers, and were similar to those who maintained follow-up with respect to defect severity type (A-D, $p=0.57$), ECMO use ($p=0.35$), number of affected organ systems ($p=0.36$), and number of providers following after discharge ($p=0.33$). Seventeen (24%) families completed the survey, of whom eight (47%) were deemed lost to follow up to specialist providers. Families reported distance and time constraints, access to CDH-specific information and care, access to CDH-specific resources, and access to healthcare as significant barriers to care. All respondents were interested in a multidisciplinary CDH clinic.

Conclusions

CDH survivors require multidisciplinary care beyond initial repair, but attrition to follow-up after discharge is high. A multidisciplinary CDH clinic may address caregivers' perceived barriers.

Keywords

Congenital diaphragmatic hernia, multidisciplinary clinic, access to healthcare, aftercare

Introduction

Congenital diaphragmatic hernia (CDH) is a congenital defect in the diaphragm, thereby allowing abdominal contents to herniate into the chest and severely impair lung and pulmonary vasculature development. This results in varying degrees of pulmonary hypoplasia and pulmonary hypertension.[1] CDH occurs in approximately 1 in 3000 live births, making this condition more common than many other congenital anomalies.[2] Overall mortality following CDH repair has slowly decreased since the mid-1990s, a trend attributed to increased prenatal diagnosis, improved ventilation strategies, and delayed surgical repair.[3,4] However, CDH contributes to ongoing morbidity following discharge, with survivors facing numerous long-term sequelae, including respiratory, gastrointestinal, neurodevelopmental, and musculoskeletal comorbidities.[4-15] Thus, CDH survivors represent a growing cohort who require complex long-term follow-up care that is traditionally coordinated by primary care providers.[16] To this end, a guideline for primary care providers on the detection and management of CDH-associated comorbidities was published in 2008 by the American Academy of Pediatrics.[16]

As an alternative to PCP-directed care, CDH patients may obtain follow-up care in a multidisciplinary CDH clinic. The multidisciplinary clinic model is increasingly employed for various other complex pediatric populations (tracheoesophageal fistula, anorectal malformations, short bowel syndrome, cystic fibrosis, etc.), where it has led to improvements in outcomes.[17-20] The first multidisciplinary CDH clinic was established at Boston Children's Hospital in 1990, and this model has since been extended to select US, Canadian, and European centers.[21-22] Despite significant practice variation across clinics, this model is considered a more holistic and consolidated form of complex follow-up care. Furthermore, clinics serve as hubs for research efforts in the area of long-term management of CDH patients.[22-23]

Still, a large portion of the CDH survivor population remains outside the encatchment of multidisciplinary CDH clinics and, thus, continues to obtain follow-up care for multiple comorbidities in a more fragmented fashion. Such is the case of the CDH patients who underwent repair at Riley Hospital for Children at IU Health, where a multidisciplinary CDH clinic has yet to be established. Thus, we sought to characterize the barriers to follow-up care that CDH patients and families face in this context. We hypothesized that survivors who underwent repair at our institution would 1) perceive significant barriers to accessing follow-up care and 2) perceive a personal benefit to a multidisciplinary clinic.

Methods

Study design and patient selection

After IRB approval (#1811277969), a retrospective review of all CDH patients who underwent repair at Riley Hospital for Children (RHC) at Indiana University Health from January 1st, 2010 through December 31st, 2019 was conducted. Waiver of consent was granted for retrospective chart review of patients, and informed verbal consent was obtained specifically for participation in the survey. Patients older than 12 years at the time of study were excluded due to insufficient medical records of those from greater than 12 years prior. Patients known to be deceased at the time of study were also excluded. The electronic medical record (EMR) was reviewed for the following data points: CDH Study Group defined defect type (A, B, C, D), repair details (primary versus patch repair, etc.), the need for extracorporeal membrane oxygenation (ECMO) support, whether the diagnosis was prenatally known, other operations, major complications during admission, discharge diagnoses, providers arranged for follow-up at discharge, current follow up status with specialty providers at our institution, and insurance status at the time of repair and at last known encounter.[3] Regarding follow-up status, patients were considered either 1) continuing or having completed recommended follow-up visits if they completed or continued follow-up with all recommended specialty providers, or 2) lost to follow-up if they were lost from any recommended specialty provider at our institution while follow-up was still being recommended.

To maximize response rate, caregivers of eligible patients were contacted twice by mail, with each mailing approximately 6 months apart. Caregivers who had not responded were then contacted a third time by phone. After verbal consent was obtained, surveys were completed by phone, mail, or email, according to caregiver preference. Each family was asked to complete a

survey regarding caregiver demographics, functional status, perceived barriers to care, quality of life as assessed by PedsQL™ scales, and perceptions of the effect of a multidisciplinary CDH clinic on their child's care.[24] All survey questions other than the PedsQL™ questions were developed specifically for this study by the research team and were not independently validated. The entirety of the survey is provided in **Supplementary Material**. Responses were recorded in a RedCap™ database.

Survey response analysis

Survey responses were studied, and common themes as related to barriers of care were extrapolated following qualitative review by two authors (NH, BG). PedsQL™ scoring was performed according to publicly available instructions on the PedsQL™ website.[25]

Statistical analysis

Descriptive statistics were used largely throughout the study, and data are presented as n (%) or median [interquartile range], where applicable. Missing or unavailable data was excluded during analysis. Bivariate analysis comparing patients who were and were not lost to follow up were used to determine if any differences existed in patient or disease characteristics. Categorical variables were compared by Fisher's exact test, and continuous nonparametric variables by Mann-Whitney U test; $p<0.05$ was statistically significant.

Results

Patient characteristics

One hundred fourteen patients were identified in the institutional database (**Figure 1**). After exclusions for deaths prior to study (n=43) and age > 12 years at time of study (n=1), 70 patients were eligible for contact. The majority underwent repair as neonates (age <30 days at repair, n=54, 77%), with a median age at repair of 5 days (IQR 3-18 days). Defects were most commonly left-sided (n=52, 74%), severity type B (n=32, 46%), and repaired primarily (n=44, 63%). Twelve patients underwent ECMO support during their course (17%) (**Table 1**).

Follow-up status

From the 70 eligible patients, thirty-seven (53%) continued to have or had graduated from regular follow-up with specialty providers at our institution, while 33 (47%) had been lost to follow-up of their CDH-related needs with providers who recommended continued follow-up for that child (**Figure 1**). There was no association between follow-up status and severity of CDH disease, according to the patient's ECMO support needs (p=0.35), defect severity type (p=0.57), primary versus patch repair (p=0.62), or age at repair (p=0.17). There were also no differences observed in regards to prenatal vs postnatal diagnosis (p=0.81). Patient insurance status at the time of repair and at last known follow-up were also no different between the groups (p=0.62 and 0.35, respectively). Furthermore, there was no association between follow-up status and complexity of comorbidities at discharge, according to the patient's number of affected organ systems (p=0.36) or number of additional providers for which follow-up was arranged at discharge (p=0.33). **(Table 1)**

Survey respondents

Of the 70 families contacted, 17 caregivers (24%) responded to the survey. Within the survey respondent subgroup, median age at the time of study was 3.7 years (IQR 2.3-6.2 years), and median age at repair was 9 days (IQR 2-18 days). Most defects were left-sided (n=13, 76%), severity type B (n=8, 47%), and repaired primarily (n=10, 59%). Five (29%) patients underwent ECMO support during their hospitalization. Nine (53%) were continuing or had graduated from follow-up with CDH-related specialists at our institution, and 8 (47%) were considered lost to follow-up with specialists who provided CDH-related care (**Figure 1**). Fourteen (82%) reported that their care was coordinated through a primary care provider, of which 5 (36%) were lost to follow-up with specialty providers at our institution. (**Table 2**).

Survey responses: barriers to care

Sixteen respondents completed portions of the survey pertaining to perceived barriers to care, with one respondent not completing these free-response questions. Barrier themes are summarized in **Figure 2**. The most reported barrier to follow-up care was physical distance and time constraints (62.5%). Median driving distance from home address to our institution was 35.1 miles (IQR 13.2-91.5 miles), and median reported travel time for appointments was 0-1 hours. Six (35%) respondents reported missing work for care or appointments. One (6%) respondent reported moving approximately 150 miles from previous residence to be closer to appointments. One (6%) respondent relied on Medicaid cabs and family/friends for transportation to appointments, and all others travelled by a personal car.

Lack of CDH-specific information and care was reported by 6 (37.5%) respondents who cited various ongoing concerns for which a specialist was preferred. These concerns included lack of coordination among specialists, uncertainty over the appropriate specialist for CDH-related questions, what to do in the event of a suspected CDH complication, need for continued

follow-up imaging, suspicion for patch-related symptoms, prognostic concerns, and the impact of COVID-19 on CDH survivors.

Limited access to resources was reported by 5 (31%) respondents, specifically citing support services (home health care, mental health, family support) and home supplies (oxygen, gastrostomy tubes) as greatest needs. Limited access to healthcare was reported by 4 (25%) respondents, citing difficulties with insurance coverage, financial strain, and access to non-CDH providers. One (6%) respondent reported no perceived barriers.

All respondents felt they would benefit from a multidisciplinary CDH clinic. Seven (24%) respondents provided further comment on this proposal, with four respondents emphasizing its potential for added convenience, and three respondents interested in easier access to specialists.

Survey responses: quality of life

PedsQL^{TM} scores are summarized in **Table 3**. There was no significant difference in overall quality of life scores between patients who were continuing follow-up or were lost to follow-up (p=0.54).

Discussion

In this single-institution retrospective review and patient survey study, roughly half of CDH repair survivors were lost to follow-up with specialty providers. Caregivers participating in the survey reported several barriers to their child's follow-up care, including time and distance constraints, access to specialists for ongoing CDH-related concerns, access to CDH-related resources and services, and general access to the healthcare system. All caregivers felt a multidisciplinary CDH clinic would positively impact their child's care.

Loss to follow-up in the CDH survivor population represents a significant barrier to long-term follow-up, and thus it may adversely impact developmental and functional outcomes. Structured monitoring for numerous respiratory, gastrointestinal, neurodevelopmental, and orthopedic comorbidities is recommended through the age of 16, regardless of initial disease severity.[16] Late surgical complications have also been reported, further emphasizing the need for long-term follow-up with a pediatric surgeon.[8] Despite these recommendations, loss to follow-up occurred at a high rate at our institution, and likely does at other institutions, though the loss rates are rarely reported in CDH literature. In our study of 70 survivors, 47% were lost to follow-up. This is in line with other studies in the literature, one of which noted 43% lost from a cohort of 40 CDH survivors.[26]

Factors associated with loss to follow-up have not been extensively studied. In this cohort, follow-up status was not associated with measures of disease severity (ECMO support, defect type, repair type) or burden of comorbidities at discharge. Conversely, these associations were identified by Takayasu et al, and it was interpreted that caregivers of children with less severe disease were more likely to discontinue follow-up due to a lower perceived need for follow-up.[26] However, this analysis was performed on a larger sample of survivors of various

congenital anomalies, of which CDH was a minority: of the 306 survivors included, only 40 had undergone CDH repair.[26] Otherwise, there is relatively little literature on long-term compliance with recommended follow-up in complex pediatric surgical patients, and further research is needed to adequately understand and address attrition.

Though the impact of the multidisciplinary clinic model on access to care has not yet been studied in the CDH survivor population, we propose it may lead to improvements in outcomes and thus merit future prospective study. Three of the barriers reported in this cohort - caregiver time and distance constraints, lack of access to CDH-specific information and care, and specialist incoordination - may be directly addressed by a multidisciplinary CDH clinic. There is evidence in pediatric primary care literature that the analogous "medical home" model may be associated with reductions in caregiver strain, days of missed work, and overall healthcare utilization in children with complex healthcare needs.[27-30] Multidisciplinary clinics are also designed to increase coordination across several specialists while providing families with a single point of contact (nurse care coordinator, case manager, etc.).[22] Thus, the multidisciplinary model may effectively address time and distance constraints, simplify caregiver access to specialists for CDH-specific information and care, and coordinate care between specialists, all of which were barriers or concerns reported here by the primary caregivers of our cohort. Though the multidisciplinary clinic model less directly addresses barriers related to resources and access to healthcare, increased access to nutritionists, psychologists, nurses, and social workers on the multidisciplinary team may facilitate referral to the appropriate support programs.

The benefits offered by multidisciplinary clinic implementation may be strongly enhanced by telemedicine capabilities. Even prior to the COVID-19 global pandemic, telemedicine has been an emerging form of care with marked potential to address the prominent

geographic disparities in pediatric surgical care delivery.[31,32] Furthermore, the benefits of appropriately implemented telemedicine services directly address the caregiver barriers reported here. In multiple studies, telemedicine protocols have demonstrated significantly reduced time and travel burden in surgical patients.[33] In a multidisciplinary context, the implementation of videoconferencing may facilitate provider coordination, relieving caregivers of the duty to coordinate and directly improving their perceptions of care.[34] Acceptability to both providers and patients of the virtual visit in place of the traditional consultation has been demonstrated in multidisciplinary clinics in both urban and rural settings.[35,36] Telemedicine consultations have been successfully utilized for speech, language, and behavioral therapy services as well.[37] Implementation of this technology is not without hurdles and must be tailored to patients' and providers' individual needs, but effective telemedicine capabilities will be an indispensable component of multidisciplinary follow-up care for CDH patients.

While this study takes a novel approach to understanding and addressing postoperative needs and barriers of CDH patients, it faces several limitations. These include its single-institution, retrospective design. The survey component may also be subject to selection bias, as caregivers with greater needs, higher motivation to contribute to research, and more positive perceptions of their child's specialty care were likely more apt to respond. Very limited conclusions can be drawn from quality of life scores due to small sample size and age variation. Lastly, the survey response rate was lower than anticipated, limiting the significance of all conclusions drawn here. Nonetheless, this study provided meaningful insight into the poor retention of this group at our institution.

Conclusions

Survivors of CDH repair require complex follow-up care for detection and management of CDH-associated comorbidities. Roughly half of survivors are lost to recommended follow-up care, and survivors face numerous barriers to accessing care. A multidisciplinary clinic approach may address these barriers.

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Figure 1: Patient selection

Exclusions, follow-up status, and survey respondent groupings. 70 eligible caregivers were contacted, with 17 total survey respondents from both follow-up status categories. Follow-up status was determined via review of the electronic medical record, independent of survey response.

Figure 2: Survey responses

Summary of survey respondents' barriers to care, including four principal themes ascertained from free responses. Seventeen responses were received in total, with 16 respondents completing the survey and one respondent omitting barrier-related survey questions. The area of each circle is scaled to its corresponding percentage. RHC, Riley Hospital for Children; CDH, congenital diaphragmatic hernia.

Supplementary material: Caregiver Survey

Complete survey sent to all eligible caregivers. All questions were free-response, except where scale is provided.

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Table 1: Demographics of Discharged CDH Patients

	All patients (n=70)	Continuing follow-up (n=37)	Lost to follow-up (n=33)	p-value
Age at repair				0.17
< 30d	54 (77%)	26 (70%)	28 (85%)	
≥ 30d	16 (23%)	11 (30%)	5 (15%)	
Sex				>0.99
M	43 (61%)	23 (62 %)	20 (61%)	
F	27 (39%)	14 (38%)	13 (39%)	
Prenatally Diagnosed				0.81
Yes	28 (40%)	14 (38%)	14 (42%)	
No	42 (60%)	23 (62%)	19 (58%)	
Defect side				0.57
Left	52 (74%)	28 (76%)	24 (73%)	
Right	16 (23%)	7 (19%)	9 (27%)	
Bilateral	1 (1%)	1 (3%)	0 (0%)	
Unknown	1 (1%)	1 (3%)	0 (0%)	
CDH Study Group defined defect type				0.57
A	18 (21%)	11 (30%)	7 (21%)	
B	32 (40%)	14 (38%)	18 (55%)	
C	18 (24%)	10 (27%)	8 (24%)	
D	3 (4%)	2 (5%)	1 (3%)	
Repair type				0.62
Primary	44 (63%)	22 (59%)	22 (67%)	
Patch	26 (37%)	15 (41%)	11 (33%)	
ECMO use	12 (17%)	8 (22%)	4 (12%)	0.35
Number of organ systems impacted at discharge				0.36
0	33 (47%)	15 (41%)	18 (55%)	
1-2	24 (34%)	15 (41%)	9 (27%)	
3+	13 (19%)	7 (19%)	6 (18%)	
Number of additional providers following after discharge				0.33
0	24 (34%)	14 (38%)	10 (30%)	
1-2	30 (43%)	16 (43%)	14 (42%)	
3+	15 (21%)	6 (16%)	9 (27%)	
Unknown	1 (1%)	1 (1%)	0 (0%)	
Insurance at Repair				0.62
Medicaid/Self Pay	44 (63%)	22 (59%)	22 (67%)	
Commercial	26 (37%)	15 (41%)	11 (33%)	
Last Active Insurance				0.35
Medicaid/Self Pay	34 (49%)	20 (54%)	14 (42%)	
Commercial	36 (51%)	17 (46%)	19 (58%)	

Additional provider counts do not include pediatric general surgery, primary care providers, or skilled nursing visits. Note: 1 patient was transferred to another center prior discharge, and thus additional provider count is unknown. IQR, interquartile range; ECMO, extracorporeal membrane oxygenation.

Table 2: Demographics of Survey Respondents

Variable	n (%)
Sex	
M	10 (59%)
F	7 (41%)
Age at time of study	
1-12 mo	2 (12%)
13-24 mo	5 (28%)
2-4 y	6 (36%)
5-7 y	2 (12%)
8-12 y	2 (12%)
Defect side	
Left	13 (76%)
Right	4 (24%)
Defect size	
A	3 (18%)
B	8 (47%)
C	5 (29%)
D	1 (6%)
ECMO	5 (29%)
Repair type	
Primary	10 (59%)
Patch	7 (41%)
Number of organ systems impacted at discharge	
0	8 (47%)
1-2	3 (18%)
3+	6 (35%)
Number of specialty providers needed for care after discharge	
0	4 (24%)
1-2	5 (29%)
3+	8 (47%)
Follow-up status	
Continuing or graduated	9 (53%)
Lost	8 (47%)
Care coordination	
Primary care provider	14 (82%)
Other provider	3 (18%)
Insurance at Repair	
Medicaid/Self Pay	8 (47%)
Commercial	9 (53%)
Last Active Insurance	
Medicaid/Self Pay	9 (53%)
Commercial	8 (47%)

Additional provider counts do not include pediatric general surgery, primary care providers, or skilled nursing visits. IQR, interquartile range; ECMO, extracorporeal membrane oxygenation

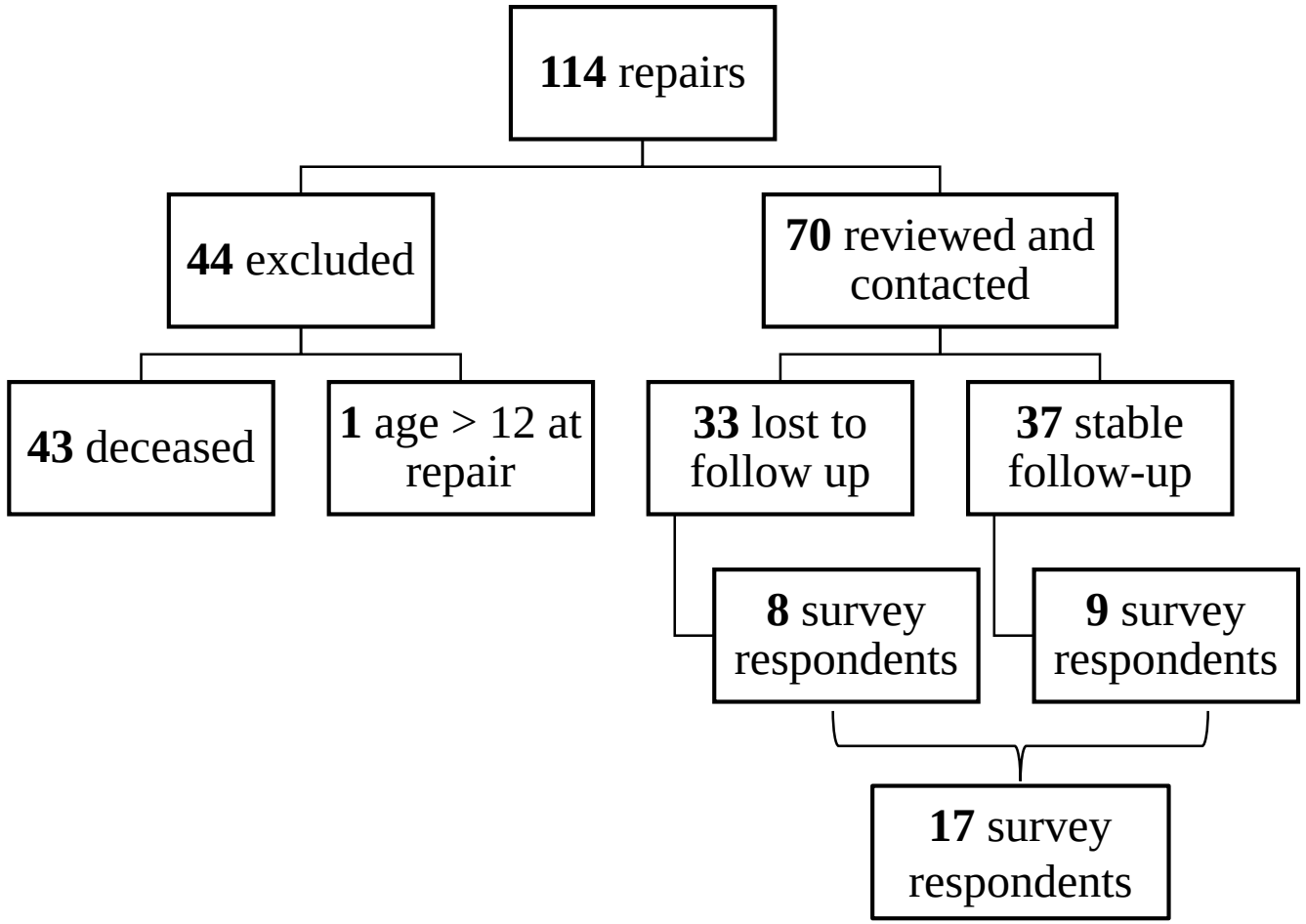
Table 3: Quality of Life in Survey Respondents

Age group	Total score	Physical functioning	Physical symptoms	Emotional functioning	Social functioning	Cognitive/school functioning
1-12 mo (n=2)	77	83	73	81	91	56
13-24 mo (n=5)	78	78	85	87	82	58
2-4 y (n=6)	84	85	-	86	82	96*
5-7 y (n=2)	86	92	-	83	93	73
8-12 y (n=2)	71	69	-	73	75	68

Means PedsQL scores by age group at the time of survey and dimension. See **Appendix** for full survey. Scores range 0-100, with higher scores correlating with better functioning or fewer symptoms. PedQL scales do not assess the "physical symptom" domain in children older than 24 mo, as indicated by dashes. Cognitive functioning is assessed prior to 24 mo, and is replaced with school functioning beyond 24 mo. * indicates that only 2/6 caregivers in this age group answered school functioning-related questions, as the remaining 4 children in this group were not yet attending school or daycare.

Figure 1

Click here to access/download;Figure;Figure 1.pptx



Responder Characteristics	**n (%)**
Continuing follow-up at RHC	15/17 (88%)
Care coordinated through primary care provider	14/17 (81%)
Barriers to Post-Discharge CDH Care	**# affected (%)**
Maintaining appropriate follow-up	33/70 (47%)
Physical proximity and time constraints to care	10/16 (62.5%)
Access to CDH specific information and care	6/16 (37.5%)
Access to resources (home aid, supplies, therapies, family support)	5/16 (31%)
Access to healthcare (insurance, financial, non-CDH providers)	4/16 (25%)
No barriers perceived	1/16 (6%)
Would benefit from a multidisciplinary CDH clinic	16/16 (100%)