



Hospitalisations of children and adults with Hypoplastic Left Heart Syndrome in Texas from 2009 to 2019

Original Article

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Abstract

Introduction: Hypoplastic Left Heart Syndrome accounts for a significant proportion of CHD morbidity and mortality, despite improvements in care and improved survival. This study evaluates number of, reasons for, and trends in discharges of patients with hypoplastic left heart syndrome over 11 years in Texas. **Methods:** The Texas Inpatient Discharge Dataset Public Use File captures almost all discharges in Texas and was reviewed from 2009 to 2019. Discharges of patients ≥ 5 years of age and diagnosis codes for Hypoplastic Left Heart Syndrome were included. The admitting and principle diagnoses were categorised and all discharges were evaluated for procedures performed. Descriptive and univariate statistical analyses were performed. **Results:** A total of 1024 discharges were identified with a 16.9% annual increase over the study period. Median length of stay was 4 [IQR: 2–8] and there were 17 (1.7%) in-hospital mortalities with no differences across age groups. Seven (17.1%) discharges of patients 25+ years were uninsured, higher than other age groups ($p < 0.001$). The most common admitting diagnosis was CHD and 224 (21.9%) of discharges included a procedure, including 23 heart transplants. Discharges occurred from 67 different hospitals with 4 (6.0%) representing 71.4% of all discharges. **Conclusions:** Discharges of Hypoplastic Left Heart Syndrome have increased rapidly, particularly in the older age groups and were spread over a large number of hospitals. Further work is needed to understand the interplay between Hypoplastic Left Heart Syndrome and other conditions and care experiences that occur within the general population, which will become more common as this population ages and grows.

Introduction

Hypoplastic Left Heart Syndrome occurs in approximately 2.6 of every 10,000 live births in the United States.¹ While risk of early mortality remains high in this complex condition, improvements in medical and surgical care have resulted in improved survival over recent decades.² The current classical three-stage palliation approach, leading to a total cavopulmonary connection (Fontan), results in significant circulatory derangements. Known long-term complications and risks for these patients include risk for arrhythmias, liver dysfunction, thromboembolism, and heart failure.^{3–6} All patients with Fontan circulation including those with Hypoplastic Left Heart Syndrome have a significant burden of hospitalisations particularly in adulthood.⁷ The long-term trajectories of this patient population are continuously evolving and continue to encompass significant uncertainty.

While previous reports have looked at trends in hospitalisations of all adults with CHD, the Hypoplastic Left Heart Syndrome population is unique with a different hospitalisation profile compared to the overall adults with CHD population.⁸ This study aims to evaluate hospitalisation characteristics and outcomes of children and adults with Hypoplastic Left Heart Syndrome as well as assess for trends in these characteristics over an 11-year span.

Methods

Data source

Data were obtained from the Texas Inpatient Discharge Dataset from 2009 to 2019.⁹ The Texas Inpatient Discharge Dataset is an administrative database that captures most discharges in the state of Texas with exception of hospitals located in a county with a population less than 35,000, or those located in a county with a population more than 35,000 and with fewer than 100 licensed hospital beds and not located in an area that is delineated as an urbanised area by

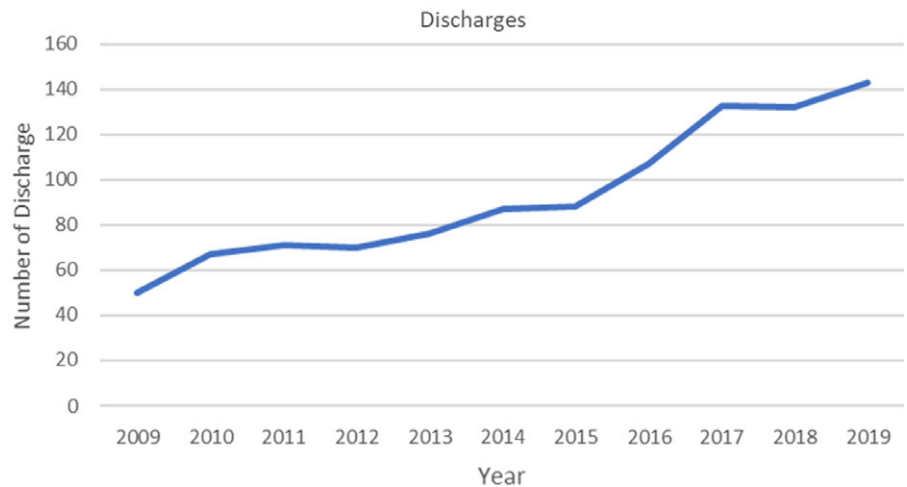


Figure 1. Number of discharges of Hypoplastic Left Heart Syndrome patient by year.

the United States Bureau of the Census. The data are collected and maintained by the Texas Department of State Health Services, Center for Health Statistics. Data are deidentified when it is submitted to the dataset from the hospitals. The Institutional Review Board for the University of Texas at Austin Dell Medical School waived the need for approval of this study as it consisted of existing, deidentified data.

Study population

The Texas Inpatient Discharge Dataset provides an admitting diagnosis, a principle diagnosis, up to 24 other diagnoses, a principle procedure and up to 24 other procedures for each hospitalisation record. From 2009 through the third quarter of 2015, diagnoses and procedures were coded using the standard International Classification of Diseases, 9th edition (ICD-9). Records from the fourth quarter of 2015–2019 were coded using the 10th edition (ICD-10).

Discharges of patients ≥ 5 years of age at discharge and diagnosis of Hypoplastic Left Heart Syndrome (ICD-9: 746.7, ICD-10: Q23.4) were included. We excluded from analysis; discharges from long-term care, mental and behavior health facilities, substance abuse centers, and unknown center type, discharges with missing information on the type of admission, sex, age, race, ethnicity, length of stay, discharge status, admitting diagnosis, or principle diagnosis, and interim entries. Additionally, discharges with an ICD-9/10 procedure codes consistent with Fontan completion (ICD-9: 39.21, ICD-10: 02160** or 02170**) in those aged 5–9 years were excluded as the focus of the analysis was post-Fontan hospitalisations.

Study outcomes

The primary aim of the study was to assess indications for hospitalisation, procedures performed, and outcomes by age and over time.

The Texas Inpatient Discharge Dataset categorises age into 16 groups. These were further collated into 5–9 years, 10–14 years, 15–19 years, 20–24 years, and 25+ years. Insurance status was grouped into private insurance, Medicare, Medicaid, uninsured, other, and unknown. Admitting and principle diagnoses were grouped into 24 categories for analysis. Procedures were identified by ICD-9 and ICD-10 procedure codes and are listed in

Supplemental Table 1. Hospitalisation outcomes included in-hospital mortality and length of stay.

Statistical analysis

Descriptive statistics were reported for demographics, hospitalisation characteristics, and outcomes. Length of stay is reported in median [interquartile range] days. All other variables are presented as proportions. Chi-square and Fisher's exact test were utilised to analyse discrete variables. Kruskal–Wallis test was utilised to analyse length of stay. Statistical analyses were performed using R and RStudio.¹⁰ All statistical tests were 2-tailed and a p-value < 0.05 was considered significant.

Results

Overall discharges

A total of 1024 eligible discharges were identified. Of these, 402 (39.3%) were of female patients, 777 (75.9%) were white, 395 (38.6%) were Hispanic, 379 (37.0%) had private insurance, and 25 (2.4%) were uninsured. Overall, 504 (49.2%) discharges were of patients between 5 and 9 years of age and 819 (80.0%) discharges were from freestanding children's hospitals. In 2009, there were 50 discharges which rose to 143 discharges in 2019 (Fig 1). This represents a 186% total increase and a 16.9% annual increase over the study period. Median length of stay was 4 [2–8] days and there were 17 (1.7%) in-hospital mortalities. Complete discharge demographics are presented in Table 1. Discharges occurred from 67 different hospitals with 4 (6.0%) hospitals representing 71.4% of all discharges while 56 (83.6%) hospitals had 10 or fewer discharges over the study period representing an average of less than 1 discharge per year over that time (Fig 2). A density map of discharges by patient location is shown in Figure 3.

Admitting and principle diagnoses

The most frequent admitting diagnoses were related to CHD with 174 (17.0%) followed by gastrointestinal diagnoses with 142 (13.9%) and pulmonary diagnoses with 102 (10.0%). The most frequent primary diagnoses were also related to CHD with 167 (16.3%) followed by infectious diagnoses with 159 (15.5%) and gastrointestinal diagnoses with 149 (14.6%). The breakdown of admitting and primary diagnosis categories are presented in

Table 1. Overall discharge demographics and outcomes.

Variable	
Total number	1024
Female, n (%)	402 (39.3)
Race, n (%)	
American Indian/Eskimo/Aleut	5 (0.5)
Asian or Pacific Islander	10 (1.0)
Black	100 (9.7)
White	777 (75.9)
Other	132 (12.9)
Hispanic, n (%)	395 (38.6)
Age, n(%)	
5–9 years	504 (49.2)
10–14 years	265 (25.9)
15–19 years	144 (14.1)
20–24 years	70 (6.8)
25+ years	41 (4.0)
Insurance, n (%)	
Uninsured	25 (2.4)
Medicaid	586 (57.2)
Medicare	2 (0.2)
Private	379 (37.0)
Other	32 (3.1)
Discharge from freestanding children’s hospital, n (%)	819 (80.0)
Length of stay (days), median [IQR]	4 [2–8]
In-hospital mortality, n (%)	17 (1.7)

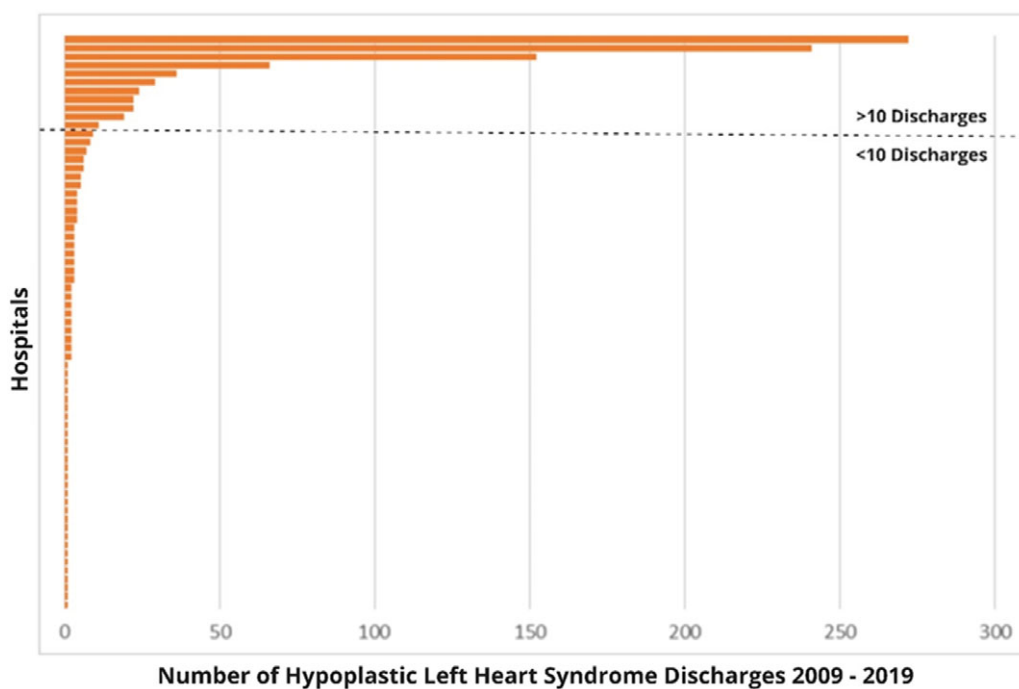


Figure 2. Number of Hypoplastic Left Heart discharges by hospital.

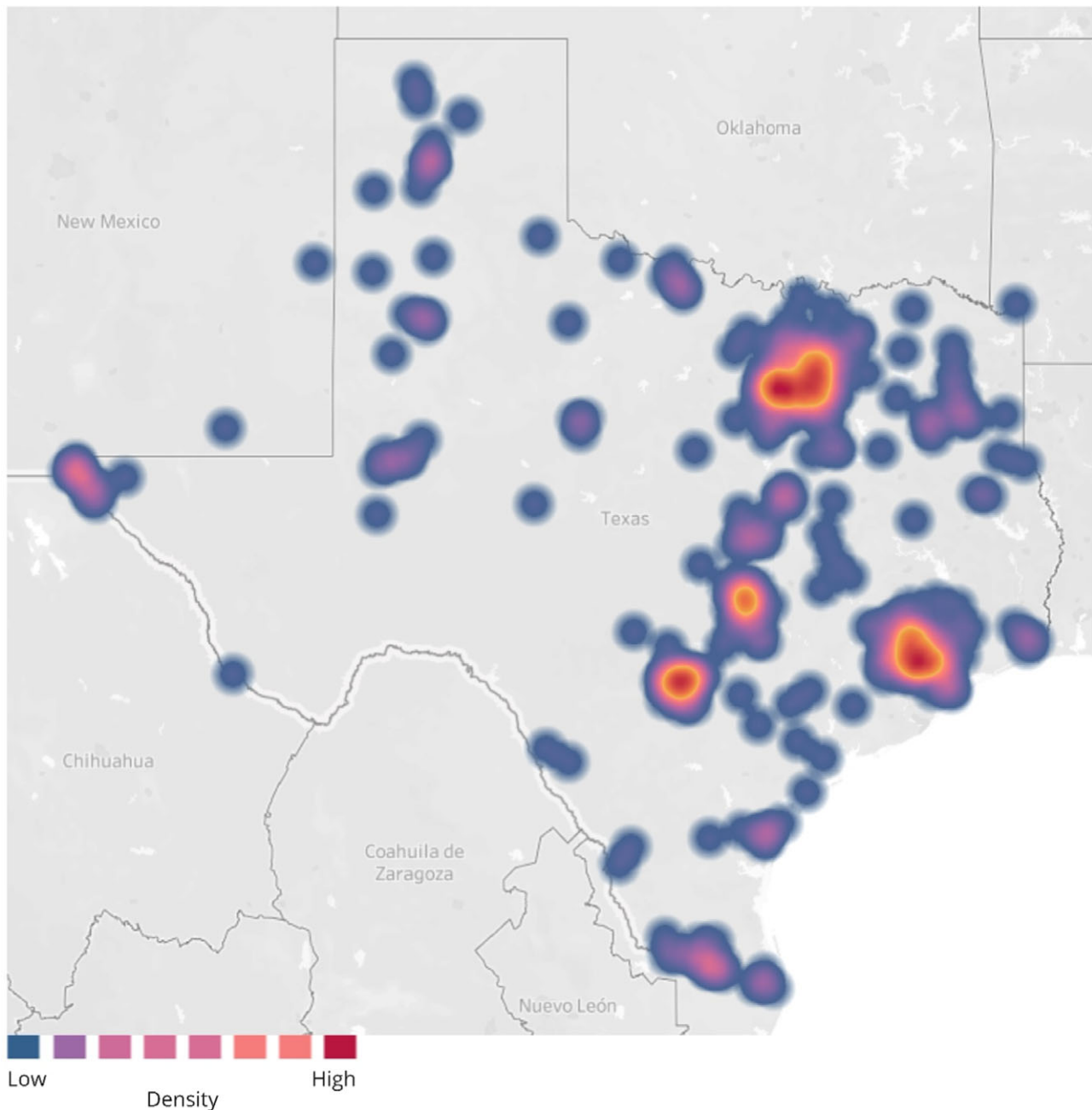


Figure 3. Density map of Hypoplastic Left Heart Syndrome discharges by location of residence.

Tables 2 and 3. The most frequent primary diagnosis for in-hospital mortalities was CHD with 4 (23.5%) followed by infection with 3 (17.6%), neurologic with 2 (11.8%), and gastrointestinal with 2 (11.8%).

Procedures performed

A total of 224 (21.9%) discharges included a surgical or catheter-based procedure. There were 23 (2.2%) heart transplants representing 10.3% of all discharges with procedures and 5 (0.5%) VAD placements representing 2.2% of all discharges with procedures. The 23 heart transplants represented 0.8% (23/3046) of all heart transplants performed in Texas over the same period.¹¹ Additionally, 26 (2.5%) discharges included a gastrointestinal

intervention and 12 (1.2%) discharges included a musculoskeletal intervention. Discharges of patients with a cardiac intervention had a longer length of stay (6 [2–12] days vs. 3 [2–6] days, $p < 0.001$) but no difference in in-hospital mortality (2.2% vs. 1.5%, $p = 0.552$).

Characteristics by ages

All age groups had an increase in numbers of discharges across the study period (Fig 4). The highest proportion of discharges throughout the entire study period was in the 5–9 year age group. However, the proportion fell from 56% of discharges in 2009 to 42.7% of discharges in 2019. This was offset by a rise in proportion of discharges in the 20–24 year age group which rose from 0% in

Table 2. Frequency of admitting diagnoses overall and by age group.

Admitting diagnosis category	Age (years)					
	All	5-9	10-14	15-19	20-24	25+
Total	1024	504	265	144	70	41
CHD	174	93	49	20	10	2
Gastrointestinal	142	61	50	21	8	2
Pulmonary	102	65	27	3	6	1
Infection	69	47	11	4	4	3
Fever	61	38	14	6	3	0
Fluid Status	56	27	20	6	2	1
Arrhythmia	53	17	13	17	5	1
Other	52	25	10	8	8	1
Neurological	40	18	8	12	2	0
Abnormal Vital Signs	38	20	13	4	1	0
Abnormal Labs	36	21	10	4	1	0
Heart Failure	31	16	6	8	1	0
Shortness of Breath	25	10	4	5	1	5
Pregnancy	24	0	0	1	8	15
Device Complications	22	13	5	1	0	3
Chest Pain	20	4	3	6	5	2
Musculoskeletal	18	6	3	8	1	0
Skin/Soft Tissue	15	7	2	3	2	1
Non-Cardiac Vascular	14	6	4	2	0	2
Mental Health	10	3	5	1	1	0
Ears Nose Throat	8	6	1	0	1	0
Trauma/Injury	7	1	5	1	0	0
Renal	5	0	1	2	0	2
Arrest	2	0	1	1	0	0

Lowest Proportion Highest Proportion

Table 3. Frequency of principle diagnoses overall and by age group.

Principle diagnosis category	Age (years)					
	All	5-9	10-14	15-19	20-24	25+
Total	1024	504	265	144	70	41
CHD	167	83	52	20	10	2
Infection	159	115	23	13	7	1
Gastrointestinal	149	52	62	22	10	3
Pulmonary	105	69	25	4	3	4
Heart Failure	64	28	18	21	1	4
Arrhythmia	60	21	14	21	3	1
Other	48	26	10	3	7	2
Neurological	41	19	9	11	2	0
Fluid Status	37	19	12	4	2	0
Device Complications	35	21	4	2	5	3
Abnormal Labs	31	17	4	8	2	0
Pregnancy	24	0	0	1	8	15
Musculoskeletal	18	5	5	8	0	0
Non-Cardiac Vascular	14	8	6	2	3	1
Mental Health	11	3	4	3	1	0
Fever	11	2	5	2	1	1
Skin/Soft Tissue	9	3	1	2	2	1
Ears Nose Throat	8	6	2	0	0	0
Trauma/Injury	8	1	6	1	0	0
Renal	7	3	1	1	0	2
Abnormal Vital Signs	6	3	1	2	0	0
Chest Pain	5	0	0	1	3	1
Shortness of Breath	1	0	1	0	0	0
Arrest	0	0	0	0	0	0

Lowest Proportion Highest Proportion

2009 and 1.5% in 2010 to 10.5% in 2019 (Fig 4). The 25+ years group was most likely to be female (n = 25 (61%)) and most likely to be uninsured (n = 7 (17.1%)). The percentage of discharges from children’s hospital decreased from >80% in all age groups 19 year and younger to 52.9% in the 20–24 year age group and 17.1% in the 25+ year group. No differences were seen across age groups for length of stay or in-hospital mortalities. (Table 4) Forty-three hospitals had at least one discharge of a patient 20 years of age or older, 2 (4.7%) centres accounted for 29.7% of the discharges, with no other centre representing more than 5.4% of the discharges (Fig 5).

Admitting diagnoses, principle diagnoses, procedures, and age

The frequency of admitting and principle diagnosis categories by age group is displayed in Tables 2 and 3. Of note, pregnancy was the most frequent admitting and principle diagnosis category for the 25+ year age group representing 36.6% of discharges. No differences were found in procedures across the age groups

(Table 5). The 20 heart transplants that occurred in patients <18 years of age constituted 5.0% (20/399) of all paediatric heart transplants in Texas during the study period.¹¹

Discussion

From 2009 through 2019 discharges of patients with Hypoplastic Left Heart Syndrome 5 years and older increased at a rapid rate and increases were seen across all evaluated age groups. This increase was coupled with an increase in proportion of discharges of older Hypoplastic Left Heart Syndrome patients, with the proportion of discharges of patients between 5 and 9 years old declining from 56 to 43% and a commensurate increase from 2 to 14% in the proportion of discharges of patients 20 years and older. Over that time CHD diagnoses were the most frequent reason for admission. More than 20% of these discharges involved an invasive procedure. Expectedly, cardiac-related surgical and catheter-based procedures represented the majority of procedures performed. In-hospital mortality was 1.7% with one of the mortalities following a heart

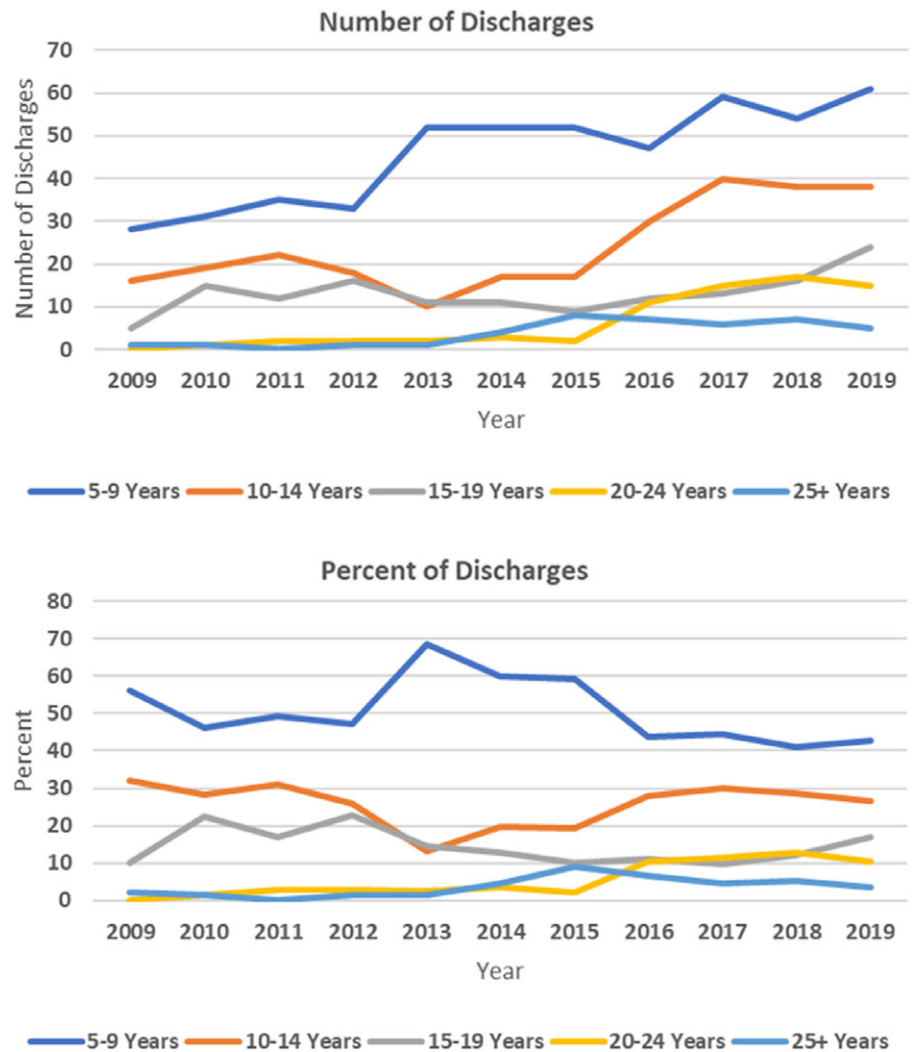


Figure 4. Hypoplastic Left Heart Syndrome discharges by age group over time.

transplant that admission. No other mortalities followed an elective cardiac procedure.

The 186% growth in discharges from 2009 through 2019 far outstrips the 17.1% growth of the Texas population over the same period of time.¹² O'Leary et al. identified a 32.8% and 87.8% growth in all paediatric and adult CHD hospitalisations, respectively, nationally from 1998 through 2010, far lower than the growth seen in in this study.¹³ The growth is also significantly more than the overall ACHD hospitalisations in Texas (41%) during the same time.⁸ This substantially larger increase likely is a product of relatively greater improvements in survival, with improved medical and surgical techniques, in patients with Hypoplastic Left Heart Syndrome compared to other CHD diagnoses. The increased proportion of discharges of adult patients with Hypoplastic Left Heart Syndrome found over the study period supports this hypothesis. However, this increase also points to a continued complexity and care need in this population.

The most frequent admitting and primary diagnoses in this population were cardiac-related. This supports the lifelong complexities of living with Hypoplastic Left Heart Syndrome. While CHD, arrhythmias, and heart failure contributed to a significant portion of principle diagnoses, other Fontan-related complications, such as cerebrovascular accident, hepatic dysfunction, and protein-losing enteropathy, were less common each with fewer

than 10 discharges represented. It should be noted, however, a significant portion of discharges was for other indications. This was also seen in the procedures performed during the hospitalisations, most were cardiac, however many involved other organ systems. As the Hypoplastic Left Heart Syndrome population continues to grow and age, it will be important to study the interaction of Hypoplastic Left Heart Syndrome and the Fontan circulation with other cardiac and non-cardiac conditions which occur within the general population. Greater understanding of these interactions will not only improve clinical care for the Hypoplastic Left Heart Syndrome population, but will also provide more clarity to individuals with Hypoplastic Left Heart Syndrome and their families as to what to expect and anticipate over time, reducing the uncertainty in the condition.

The complexity of Hypoplastic Left Heart Syndrome results in an increased healthcare resource utilisation particularly prior to Fontan completion.^{14,15} Despite an inability to track patients over time in this dataset, the increased resource utilisation is alluded to. The Hypoplastic Left Heart Syndrome population represents approximately 2–3% of all CHD births.¹⁶ However, in this study, the 5–17-year-old Hypoplastic Left Heart Syndrome population accounted for 5% of all paediatric heart transplants and 7% (20/284) of heart transplants of patients 1–17 years of age. This percentage disparity is likely larger as the 2–3% proportion is at birth,

Table 4. Demographics and outcomes by age group.

Variable	5–9 years	10–14 years	15–19 years	20–24 years	25+ Years	Sig.
Total number	504	265	144	70	41	
Female, n (%)	199 (39.4)	120 (45.3)	35 (24.3)	23 (32.9)	25 (61.0)	p < 0.001
Race, n (%)						
American Indian/Eskimo/Aleut	3 (0.6)	2 (0.7)	0	0	0	p = 0.008
Asian or Pacific Islander	6 (1.2)	1 (0.4)	0	1 (1.4)	2 (4.9)	
Black	36 (7.1)	36 (13.6)	17 (11.8)	8 (11.4)	3 (7.3)	
White	378 (75.0)	201 (75.8)	110 (76.4)	59 (84.3)	29 (70.7)	
Other	81 (16.1)	25 (9.4)	17 (11.8)	2 (2.9)	7 (17.1)	
Hispanic, n (%)	208 (41.3)	109 (41.1)	46 (31.9)	17 (24.3)	15 (36.6)	p = 0.026
Insurance, n (%)						
Uninsured	7 (1.4)	4 (1.5)	3 (2.1)	4 (5.7)	7 (17.1)	p < 0.001
Medicaid	308 (61.1)	158 (59.6)	78 (54.2)	27 (38.6)	15 (36.6)	
Medicare	0	0	0	1 (1.4)	1 (2.4)	
Private	173 (34.3)	96 (36.2)	56 (38.9)	32 (45.7)	13 (31.7)	
Other	15 (3.0)	6 (2.3)	4 (2.8)	4 (5.7)	3 (7.3)	
Discharge from freestanding children’s hospital, n (%)	421 (83.5)	225 (84.9)	129 (89.6)	37 (52.9)	7 (17.1)	p < 0.001
Length of stay (days), median [IQR]	4 [2–8]	4 [2–9]	4 [2–7]	3 [2–6]	3 [2–4]	p = 0.272
In-hospital mortality, n (%)	9 (1.8)	5 (1.9)	2 (1.4)	1 (1.4)	0	p = 1

Bold value represents p < 0.05.

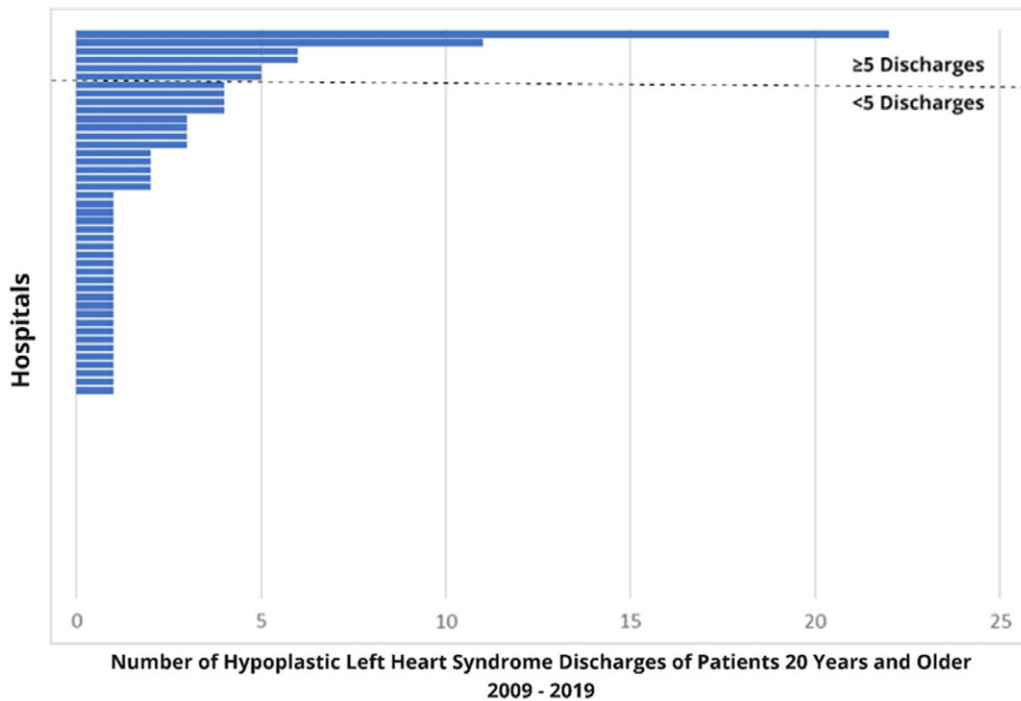


Figure 5. Number of Hypoplastic Left Heart Discharges of patients 20 years and older by hospital.

where there is significant attrition over time with a 59–64% survival at 6 years of age.¹⁷ Overall, this resource utilisation was focussed on hospitals which represented more than 70% of the discharges. However, 12.7% (n = 130) of discharges were from 1 of 56 hospitals with fewer than 1 Hypoplastic Left Heart Syndrome

discharge per year and 2.8% (n = 29) discharged came from hospitals with a single Hypoplastic Left Heart Syndrome discharge over the study period. This dispersion of resource utilisation is magnified in adults with Hypoplastic Left Heart Syndrome with only two hospitals having at least an average of one adult

Table 5. Interventions by age group.

Variable, n (%)	5–9 years	10–14 years	15–19 years	20–24 years	25+ Years	Sig.
Total number of discharges	504	265	144	70	41	
Any cardiac intervention	112 (22.2)	65 (24.5)	29 (20.1)	12 (17.1)	6 (14.6)	$p = 0.470$
Any cardiac catheterisation	82 (16.3)	46 (17.3)	21 (14.6)	8 (11.4)	2 (4.9)	$p = 0.246$
Cardiac catheter intervention	34 (6.7)	21 (7.9)	9 (6.3)	3 (4.3)	0	$p = 0.378$
Cardiac surgery	46 (9.1)	40 (15.1)	12 (8.3)	7 (10.0)	4 (9.8)	$p = 0.128$
Heart transplant	9 (1.8)	8 (3.0)	4 (2.8)	2 (2.9)	0	$p = 0.662$
Ventricular assist device	1 (0.2)	2 (0.8)	2 (1.4)	0	0	$p = 0.271$
Gastrointestinal intervention	16 (3.2)	6 (2.3)	1 (0.7)	2 (2.9)	1 (2.4)	$p = 0.520$
Musculoskeletal intervention	3 (0.6)	5 (1.9)	4 (2.8)	0	0	$p = 0.156$

Hypoplastic Left Heart Syndrome discharge per year. Additionally, 24 (20.5%) of the adult Hypoplastic Left Heart Syndrome discharges came from a hospital with only a single adult Hypoplastic Left Heart Syndrome discharge over the study period. This dispersion illustrates the need for robust mechanisms by which to care for this population and allow timely access to specialists familiar with Hypoplastic Left Heart Syndrome management across far-flung hospitals.

As patients with Hypoplastic Left Heart Syndrome require life-long medical follow-up, access to adequate health insurance is imperative. A significant transition point for health insurance is at the age of 26 years when children can no longer be covered as dependents under parental insurance. In the general population, it is estimated that 2.5–5.3% of individuals lose coverage when they turn 26 years old with resultant decreases in outpatient physician and dental visits.¹⁸ The current study revealed relatively low rates of discharges of uninsured patients for all age groups aside from the 25 years and older group with 17.1% being uninsured. This rate of uninsured discharges is significantly higher than the 6% rate of uninsured in all forms of adults with CHD discharges in a national sample.¹⁹ Further work on understanding the full impact of the lack of insurance, along with the development of solutions to prevent this situation, is urgently needed.

Limitations

The limitations of this study should be noted. As a retrospective analysis of an administrative dataset, there are the usual limitations including the lack of complete clinical data for individual patients. Further, as the unit of analysis is a hospital discharge without any unique patient identifiers, there is a possibility that a single patient may be represented multiple times if they had multiple hospitalisations during the study period.

There is the potential for misclassification of admitting and principle diagnoses as these are reliant on accurate coding of the actual diagnoses at the reporting hospitals. This risk is reduced by grouping these diagnoses into larger categories for analysis, as it is less likely the diagnoses would be misclassified into the wrong organ system. It is further possible that discharges with Hypoplastic Left Heart Syndrome diagnoses were not of true Hypoplastic Left Heart Syndrome patients and conversely, that some discharges of Hypoplastic Left Heart Syndrome patients were not identified as the Hypoplastic Left Heart Syndrome diagnosis was not recorded. This is likely a small risk given that Hypoplastic Left Heart

Syndrome has a unique diagnosis code and it is a condition that is quite unique and unlikely to be confused with others.

This analysis only captures patients who are discharged from a hospital and is unable to assess the experience of these patients outside of the hospital setting or to assess the experiences of those who have not been hospitalised during the study period.

Conclusion

Discharges of children and adults with Hypoplastic Left Heart Syndrome continue to rise at a rapid pace, particularly in the adult age groups. While many of these hospitalisations are secondary to cardiac diagnoses and procedures, this patient population is also impacted by many other conditions, similar to the general population. Further, the rates of uninsured Hypoplastic Left Heart Syndrome adults surpass the rates in general adults with CHD hospitalisations and the general population. This rate of uninsured is likely exacerbated by the loss of parental insurance coverage at 26 coupled with their underlying complex CHD potentially limiting employment opportunities. Further work must be done to understand the interplay between Hypoplastic Left Heart Syndrome and other conditions and experiences that occur within the general population. These conditions and experiences will become more common as the Hypoplastic Left Heart Syndrome population ages and grows in size. As the Hypoplastic Left Heart Syndrome population evolves, it provides a unique opportunity to develop creative solutions to the challenges faced by these patients and their families.

Supplementary material. To view supplementary material for this article, please visit <https://doi.org/10.1017/S1047951122003146>

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Conflicts of interest. None.

Ethical standards. The Institutional Review Board of the University of Texas at Austin Dell Medical School deemed this as not human subjects research as the study involved de-identified, publicly available data.

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