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Fragile X Syndrome-Associated Emergency Department Visits in the United States, 2006–2011

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Abstract

Using national data, we examined emergency department (ED) encounters during 2006–2011 for which a diagnosis code for fragile X syndrome (FXS) was present (n = 7,217). Almost half of ED visits coded for FXS resulted in hospitalization, which is much higher than for ED visits not coded for FXS. ED visits among females coded for FXS were slightly more likely to result in hospitalization. These findings underscore the importance of surveillance systems that could accurately identify individuals with FXS, track healthcare utilization and co-occurring conditions, and monitor quality of care in order to improve care and reduce FXS-associated morbidity.

Keywords

Fragile X syndrome; emergency department visits; hospitalizations; healthcare utilization

Introduction

Fragile X syndrome (FXS) is an inherited condition that can cause intellectual disability (ID), behavioral and social problems, and neurological problems (Bagni, Tassone, Neri, & Hagerman, 2012; Gallagher & Hallahan, 2012; Nazareth et al., 2016; Raspa, Wheeler, & Riley, 2017; Vekeman et al., 2015). Limited literature exists on healthcare use and expenditures of individuals living with FXS (McDermott et al., 2015; Nazareth et al., 2016; Sacco, Capkun-Niggli, Zhang, & Jose, 2013; Vekeman et al., 2015). Visits to the emergency department (ED) may highlight health problems that have not been adequately addressed or managed through standard medical care for persons living with FXS. Serious health problems may be more likely to result in hospitalizations.

Methods

In this cross-sectional analysis, we used 2006–2011 discharge data from the Nationwide Emergency Department Sample (NEDS), a product of the Healthcare Cost and Utilization Project (HCUP), Agency for Healthcare Research and Quality (AHRQ), to identify ED

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encounters for which the *International Classification of Diseases, 9th Revision, Clinical Modification* (*ICD-9-CM*) diagnosis code 759.83 for FXS was indicated in any of the 15 diagnosis fields. NEDS is the largest all-payer U.S. ED database and accounts for approximately 20% of U.S. hospital-based ED visits.

NEDS contains event-level records, not patient-level records. The HCUP databases contain no direct patient identifiers for patient-level analysis, and are consistent with the definition of "limited data sets" under the Health Insurance Portability and Accountability Act (HIPAA) Privacy Rule (U.S. Department of Health and Human Services, 2015). Because the unit of analysis is the ED encounter, a person seen in the ED multiple times in 1 year would be counted each time as a separate encounter (Weiss, Wier, Stocks, & Blanchard, 2014).

Discharge status reflects the disposition of the patient at discharge from the ED, and includes the following categories: admission to the same hospital, treated and released (stabilized in the ED and discharged home), transferred to another hospital, died in the ED before discharge, or any other disposition (Weiss et al., 2014). We categorized inpatient admission status as yes/no, with the latter referring to nonadmission ED visits where the patient was treated and released, transferred to another hospital, died in ED before discharge, or destination was unknown.

The variables in this analysis included age, age group, admission status, admission day of the week, diagnosis position of the FXS code, hospital region, hospital teaching status, median household income, primary payer, and trauma center status (trauma center/ nontrauma center). We defined admission day of the week as admission on a weekend (admitted Saturday or Sunday: yes/no). We defined hospital region according to the U.S. Census Bureau (Northeast, Midwest, South, and West). We categorized hospital teaching status as metropolitan nonteaching, metropolitan teaching, and nonmetropolitan.

We analyzed NEDS data using HCUP sample weights. Because of the complex sample design, we applied stratum weights to discharges based on the year of the discharge in order to obtain nationally representative estimates. We grouped ED visits according to gender and discharge status. We performed all statistical analyses using SAS Version 9.4 (SAS Institute Inc., Cary, NC). We used descriptive statistics (frequency counts and percentages) to compare ED visits by admission status (outcome of interest) and gender.

Results

From January 2006 to December 2011, an estimated 7,217 ED visits with a FXS diagnosis code occurred in the United States (2,698 ED visits among children aged ≤17 years and 4,519 ED visits among adults aged ≥18 years). Table 1 summarizes key characteristics of ED visits by gender. Approximately 45% (n = 3,246) of ED visits with a FXS diagnosis code were admitted to a hospital (either to the same hospital as the visit or a different hospital). Nearly one-third of children were admitted (28.2%), and slightly more than half of adults were admitted (55.0%; data not shown). Nineteen (0.3%) ED visits ended in death. ED visits with an FXS diagnosis code were predominantly among males (85.0%) and patients with either Medicaid (39.3%) or Medicare (34.8%) as the primary payer, and most

often took place in a metropolitan teaching hospital (46.2%) or a hospital with a trauma center (56.5%).

ED visits from female patients resulted in more frequent inpatient admissions than visits from male patients (51.4% versus 43.8%; Table 1). For both genders, the highest percentage of ED visits had Medicaid as the primary payer (39.3%). A small percentage had missing income information (2.2% homeless or foreign, $n = 159$). A higher percentage of males lived in zip codes with low median household incomes than that of females (1st and 2nd quartiles combined: 50.2% versus 43.1%).

Table 2 compares ED visits with an FXS diagnosis code by discharge status and gender. Males whose ED visits resulted in inpatient admissions were older than the females admitted (mean age: 40.0 years versus 36.0 years). The opposite was found for nonadmissions: females whose ED visit resulted in nonadmissions were older than their male counterparts (mean age: 26.3 years versus 24.1 years). For both genders, inpatient admission rates were higher among persons aged ≥ 65 years, in the Northeast, and those who had higher median income (4th quartile) or Medicare as the primary payer. Inpatient admissions differed by hospital teaching status and trauma center status. The inpatient admission rate was higher for ED visits by females that took place in nonmetropolitan hospitals or hospitals with nontrauma status. Regarding ED visits by males, higher inpatient admission rates occurred for metropolitan teaching hospitals or hospitals with trauma status.

Discussion

Using a nationally representative U.S. sample of ED visits, we estimated that 7,217 ED visits with an FXS diagnosis code occurred during 2006–2011. Nationally, about 1 in 7 ED visits ended in admission to the same hospital, and 6 in 7 ended in discharge home or to a different hospital (62 and 359 per 1,000 population, respectively) in 2011 (Weiss et al., 2014). In comparison, almost half (45.0%) of ED visits with a FXS diagnosis code resulted in admission to the same hospital. One study used administrative data to describe ED visits and hospitalizations among young persons with FXS in South Carolina, but did not report discharge information for ED visits (McDermott et al., 2015). Studies have reported that ED visits associated with certain chronic conditions, such as sickle cell disease, likewise result in inpatient admissions more frequently than in the general population (Dupervil, Grosse, Burnett, & Parker, 2016), whereas ED visits among children with autism spectrum disorder are less likely to result in hospital admissions (Deavenport-Saman, Lu, Smith, & Yin, 2016).

A slightly lower percentage of ED visits among females in the general population resulted in admissions than for males (14.4% versus 15.2%; Weiss et al., 2014). In contrast, ED visits among those with FXS were more likely to result in inpatient admissions among females in our study, 51.4% of ED visits compared with 43.8% among males with FXS.

This study had limitations. First, this study was restricted to a 759.83 *ICD-9-CM* code recorded in ED visits. Accuracy of the coding may be dependent on discharge status. Visits among individuals with that *ICD-9-CM* code recorded in other settings were also likely missed. Second, ED visits for individuals with FXS for whom the *ICD-9-CM* code was

never recorded were by definition not represented in this data set. Third, there could be reasons why the FXS code was used other than for a diagnosis of FXS. We may have overestimated the number of ED encounters related to FXS due to the inability to distinguish if the code was used to indicate carrier or diagnosis status. FXS tests can be ordered during the ED visit or the patient may have a premutation. In particular, an ED visit involving pregnancy, childbirth, or puerperium may indicate an *ICD-9-CM* code for FXS because a woman knows she is a premutation carrier or has full mutation FXS. Therefore, NEDS data cannot be used to determine if the ED visit is related to an FXS diagnosis or phenotype. Fourth, the number of unique patients who presented with a diagnosis of FXS cannot be determined because NEDS does not identify individuals who had more than one ED visit during the study period. Fifth, the data set does not represent all states (e.g., Texas) and does not include information from federal or military hospitals.

Conclusion

We found that almost half of ED visits with FXS diagnosis codes resulted in hospitalizations. Further research is needed to identify potentially modifiable factors that influence the high rate of inpatient admission from the ED among persons with FXS. These findings underscore the importance of surveillance systems that could accurately identify individuals with FXS, track healthcare utilization and co-occurring conditions, and monitor quality of care in order to improve care and reduce FXS-associated morbidity.

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Healthcare Cost and Utilization Project (HCUP) databases bring together the data collection efforts of state data organizations, hospital associations, and private data organizations (HCUP Data Partners) to create a national information resource of encounter-level health care data. HCUP would not be possible without their contributions. A complete listing of the state organizations can be accessed at www.hcup-us.ahrq.gov/hcupdatapartners.jsp.

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Table 1

Descriptive Characteristics of Weighted Emergency Department (ED) Visits With Fragile X Syndrome Diagnosis Codes in the Healthcare Cost and Utilization Project (HCUP) National Emergency Department Sample (NEDS), 2006–2011

Characteristic	ED Visits Among Males *n* (%)	ED Visits Among Females *n* (%)	Total ED Visits *n* (%)
Number of Visits (Row %)	6,135 (85.0)	1,083 (15.0)	7,217 (100.0)
Mean Age in Years (Standard Error)	31.0(1.0)	31.2 (1.6)	31.1 (0.9)
Age Categories			
≤5	657 (10.7)	122(11.2)	779 (10.8)
6–13	1,224 (20.0)	102 (9.4)	1,326 (18.4)
14–17	541 (8.8)	53 (4.9)	593 (8.2)
18–24	722 (11.8)	208 (19.2)	931 (12.9)
25–44	1,179(19.2)	326(30.1)	1,505 (20.9)
45–64	1,075 (17.5)	177(16.4)	1,252 (17.3)
≥65	737(12.0)	95 (8.8)	832 (11.5)
Discharge Status			
Treated and released or transferred to another hospital	3,378 (55.1)	514(47.5)	3,892 (54.0)
Admitted to hospital	2,689 (43.8)	557 (51.4)	3,246 (45.0)
Died in ED	N/S	N/S	19 (0.3)
Destination unknown	48 (0.8)	12(1.1)	60 (0.8)
Weekend Day	1,655 (27.0)	339 (31.3)	1,994 (27.6)
Hospital Region			
Northeast	1,442 (23.5)	207(19.1)	1,648 (22.8)
Midwest	1,860 (30.3)	354 (32.7)	2,214 (30.7)
South	1,828 (29.8)	344 (31.7)	2,172 (30.1)
West	1,005 (16.4)	178 (16.5)	1,183 (16.4)
Hospital Teaching Status			
Metropolitan nonteaching	2,433 (39.7)	367 (33.9)	2,800 (38.8)
Metropolitan teaching	2,747 (44.8)	588 (54.3)	3,335 (46.2)
Nonmetropolitan	954 (15.6)	128(11.8)	1,082 (15.0)
Median Household Income for Patient Zip Code			
Missing	N/S	N/S	159 (2.2)
1st quartile (lowest income quartile)	1,433 (23.4%)	217(20.0%)	1,650 (22.9%)
2nd quartile	1,642 (26.8%)	250(23.1%)	1,892 (26.2%)
3rd quartile	1,433 (23.4%)	379 (35.0%)	1,812 (25.1%)
4th quartile (highest income quartile)	1,476 (24.1%)	227(21.0%)	1,703 (23.6%)
Primary Payer			
Medicare	2,222 (36.2%)	290 (26.8%	2,512 (34.8%)
Medicaid	2,417 (39.4%)	417(38.5%)	2,834 (39.3%)
Private Insurance	1,205 (19.6%)	314 (29.0%)	1,520 (21.1%)
Other	285 (4.6%)	58 (5.4%)	343 (4.8%)

Characteristic	ED Visits Among Males *n* (%)	ED Visits Among Females *n* (%)	Total ED Visits *n* (%)
Trauma Center Status			
Trauma center	3,405 (55.5)	673 (62.2)	4,079 (56.5)
Non trauma center	2,729 (44.5)	409 (37.8)	3,138 (43.5)

Note. N/S = not shown; it indicates cell size of ≤10 or masked to not show small cell size, and is not provided per HCUP Data Use Agreement.

Table 2

Selected Characteristics of ED Inpatient Admissions and Other Discharge Status With Fragile X Syndrome Diagnosis Codes in the Healthcare Cost and Utilization Project (HCUP) National Emergency Department Sample (NEDS), 2006–2011

	ED Visits Among Males			ED Visits Among Females		
	Inpatient Admissions *n* = 2,689	Other Discharge Status[a] *n* = 3,446	Admission Rate	Inpatient Admissions *n* = 557	Other Discharge Statusa *n* = 526	Admission Rate
Characteristic	***n* (%)**	***n* (%)**	**(%)**	***n* (%)**	***n* (%)**	**(%)**
Mean Age in Years (Standard Error)	40.0 (1.4)	24.1 (1.0)		36.0(1.7)	26.3 (1.8)	
Age Categories						
≤5	200 (7.5)	457 (13.3)	30.4	32 (5.7)	90(17.1)	26.2
6–13	321 (11.9)	903 (26.2)	26.2	40 (7.2)	62(11.8)	39.2
14–17	142 (5.3%)	398 (11.6)	26.3	26 (6.4)	27(5.1)	49.1
18–24	249 (9.2%)	473 (13.7)	34.5	78 (14.0)	130(24.8)	37.5
25–44	591 (22.0)	589 (17.1)	50.1	206 (37.0)	120(22.8)	63.2
45–64	651 (24.2)	423 (12.3)	60.6	111 (20.0)	66(12.6)	62.7
≥65	534 (19.9)	202 (5.9)	72.5	65 (11.6)	30(5.8)	68.4
Weekend Day	760 (28.3)	895 (26.0)	45.9	167 (30.0)	172 (32.7)	49.3
Died in the Hospital	38 (1.4)	N/S		N/S	N/S	
Hospital Region						
Northeast	805 (29.9)	636 (18.5)	55.9	124 (22.3)	83 (15.7)	59.9
Midwest	682 (25.4)	1,178 (34.2)	36.7	197 (35.3)	157(29.9)	55.6
South	821 (30.5)	1,007 (29.2)	44.9	154 (27.7)	190 (36.1)	44.8
West	381 (14.2)	625 (18.1)	37.9	82(14.7)	96(18.3)	46.1
Hospital Teaching Status						
Metropolitan non teaching	1,012 (37.6)	1,421 (41.2)	41.6	138 (24.8)	229 (43.4)	37.6
Metropolitan teaching	1,395 (51.9)	1,352 (39.2)	50.8	330 (59.2)	258 (49.1)	56.1
Nonmetropolitan	281 (10.5)	673 (19.5)	25.3	89(15.9)	39 (7.5)	69.5
Median Household Income for Patient Zip Code						
Missing	62 (2.3)	89 (2.6)	69.7	N/S	N/S	55.6
1st quartile	625 (23.2%)	808 (23.4%)	43.6%	120 (21.5%)	97(18.4)	55.3
2nd quartile	645 (24.0%)	997 (28.9%)	39.3%	98(17.6)	152 (28.9)	39.2
3rd quartile	612 (22.8%)	821 (23.8%)	42.7%	198 (35.5%)	181 (34.4)	52.2
4th quartile	745 (27.7%)	731 (21.2%)	50.5%	136 (24.4%)	92(17.5)	59.6
Primary Payer						
Medicare	1,381 (51.4)	841 (24.4)	62.2	196 (35.3)	94(17.9)	67.6
Medicaid	765 (28.5)	1,652 (48.0)	31.7	197 (35.4)	220(42.1)	47.2
Private Insurance	469 (17.4)	737 (21.4)	38.9	148 (26.5)	167(31.9)	47.0

	ED Visits Among Males			ED Visits Among Females		
	Inpatient Admissions *n* = 2,689	**Other Discharge Status[a] *n* = 3,446**	**Admission Rate**	**Inpatient Admissions *n* = 557**	**Other Discharge Statusa *n* = 526**	**Admission Rate**
Characteristic	***n* (%)**	***n* (%)**	**(%)**	***n* (%)**	***n* (%)**	**(%)**
Other	74 (2.8)	211 (6.1)	26.0	16(2.8)	42 (8.1)	27.6
Trauma Center Status						
Trauma center	1,576 (58.6)	1,829 (53.1)	46.3	337 (60.4)	337 (64.0)	50.0
Nontrauma center	1,113 (41.4)	1,616 (46.9)	40.8	220 (39.6)	189(36.0)	53.8

Note. N/S, not shown indicates cell size of ≤10 or masked to not show small size, and is not provided per HCUP Data Use Agreement. ED = emergency department.

[a]Other discharge status includes ED visits that did not result in hospitalizations (i.e., treated and released, transferred to another hospital, died in ED or destination unknown).