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Research Paper

Preventive care services and health behaviors in children with fragile X syndrome

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ABSTRACT

Background: This is the first description of preventive care services specifically received by children and young adults with fragile X syndrome (FXS). We compare these rates to those of other pediatric populations and identify care disparities within our cohort.
Objective: Describe the frequency of preventive care services and health behaviors by young people with FXS, and identify disparities in care.
Methods: We assessed four preventive care outcomes and the total number of preventive care guidelines met among individuals under 21 years from the ongoing Fragile X Online Registry with Accessible Research Database (N = 406) using data from 2012 to 2015. We used adjusted odds ratios (AORs) from multiple logistic regression models to describe associations between demographic factors and preventive care outcomes.
Results: Seventy-five percent of our sample met dental care guidelines, 55.4% met influenza vaccination guidelines, 92.1% met immunization guidelines, and 24.4% met physical activity (PA) guidelines. Compared to children six to 10 years, younger children were less likely to have seen a dentist as recommended (AOR: 0.26) and young adults aged 16–20 were less likely to have received immunizations (AOR: 0.14) or to have engaged in recommended PA (AOR: 0.29). Black participants (AOR: 0.25) were less likely to have received an influenza vaccination than white participants. Individuals with autism (AOR: 0.25) were less likely to have sufficient PA, while individuals with hypersensitivity were more likely to have sufficient PA (AOR: 2.37) than unaffected individuals.
Conclusions: The proportion of young people with FXS that meet basic recommendations in preventive care guidelines varies according to health condition and demographic characteristics. This proportion could be increased for some groups, particularly in the cases of influenza vaccination and physical activity.

Fragile X syndrome (FXS) is the most common known cause of inherited intellectual disability (ID), affecting approximately one in 4,000–5,000 males and one in 6,000–8,000 females in the general population.[1] Children with this X-linked condition may experience developmental delays, intellectual disability or learning disabilities, and social and behavior problems.[1] Intellectual disability occurs uniformly in males with FXS, with generally more severe

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impairment seen in males than in females.[2] Although persons with developmental disabilities can receive a great deal of regular and specialized health care, there is evidence that they may receive inadequate preventive care services.[3–6] A sizable number of children with a developmental disability have an unmet health need,[7–9] and their rates of dental care,[10–12] full immunization,[6,13,14] and sufficient physical activity (PA)[15,16] are low or significantly lower than those of children without special health care needs. The use of preventive care has not been specifically studied among youth with FXS.

Preventive care has several different components including medical care, screening, and education that help adults and children lead healthier lives. There are specific guidelines for children to help ensure proper growth and development, as well as to prevent disease. The American Academy of Pediatric Dentistry (AAPD) recommends that infants begin seeing a dentist at the time of their first tooth eruption or by one year of age, followed by regular appointments every six months, to prevent dental caries and other problems.[17] CDC recommends 15 vaccines of varying doses be administered between birth and 18 years of age,[18] including the annual influenza vaccinations for all persons six months of age or older.[19] Finally, the Department of Health and Human Services (DHHS) recommends that individuals six to 17 years of age participate in 60 min of aerobic, muscle-strengthening, and bone-strengthening PA every day, while adults need approximately 150 min of PA each week.[20]

In this study, we examined a cohort of 406 children and young adults with FXS to describe their use of preventive care services and health behaviors. Our objective was to compare the frequencies of dental visits, immunizations, and PA to those recommended by current guidelines. We also tested the association of these frequencies with key demographic and health-related variables to identify disparities in care. This is the first description of preventive care utilization specifically within the pediatric fragile X community.

Methods

Data for this analysis were derived from version two of the Fragile X Online Registry with Accessible Research Database (FORWARD), a multisite observational study which began collecting baseline data in 2012 from 25 fragile X clinics across the United States. FORWARD has two primary components: the registry and the longitudinal database. The registry form collects demographic data, and is open to individuals with FXS and their family members. The longitudinal database includes a parent form, a clinician form, and three Standardized Behavioral Assessments, and is only open to those with full mutation fragile X. The parent form, clinician form, and Aberrant Behavior Checklist–Community[21] are completed annually, while the Social Responsiveness Scale, Second Edition,[22] and the Social Communication Questionnaire[23] are completed once. The registry and longitudinal components are open to individuals of any age. We ran our analyses using baseline data from the registry, parent form, and clinician form from 2012 to 2015. The informed consent or assent process varies by clinic, and the study was approved by each clinic's IRB. Additional information regarding the methodology of FORWARD can be found in Sherman et al.[24]

In our analysis we used the five preventive care and health behavior questions from the FORWARD parent form as our outcome measures: 1) time since last dental visit; 2) time since last influenza immunization; 3) level of PA; 4) immunization status; and 5) having a regular pediatrician or doctor. The form asked parents how long it had been since their child last had a dental visit or a flu shot. To match the response categories with the guidelines described in the introduction, we dichotomized them as “meets guidelines” (coded as one) if they selected “within the past year” or “does not meet guidelines” (coded as zero) if it was not in the past year. As we did not have information on biannual dental visits recommended by the AAPD,[17] nor the reason for the visit (preventive or otherwise), we assumed all participants who visited their dentist within the past year received recommended dental care, while those who last saw a dentist over one year prior did not.

DHHS guidelines recommend either 60 min of PA every day or 150 min each week, depending on age.[20] The FORWARD form asked parents how many of the past seven days the child exercised for at least 20 min. Because PA was measured in a manner what was not consistent with DHHS guidelines, we chose to use the highest amount of PA on the form, at least 20 min of daily exercise five to seven days in the past week, as a proxy for meeting PA recommendations. We excluded children under six years of age from the PA analysis as the guidelines do not apply to them.

We intended to use having a regular pediatrician or doctor as an additional outcome measure, however virtually all parents reported their child had a regular pediatrician or doctor. Our final outcome variable was categorized as meeting guidelines if the parent reported that their child was not missing any immunizations. We identified a wording error in the immunization response category “No, but do not plan to get missing immunizations,” which indicates that the person is not missing any immunizations, and also that they do not plan to make up missing immunizations. We excluded eight participants who selected this response category who would otherwise have been included in our sample.

Using the number of guidelines an individual met, we created a preventive score system as an exploratory outcome variable. If a participant met the recommendation for a preventive measure they scored a value of one, otherwise the score had a value of zero. Since there was little variation in having a regular doctor, we did not include this variable in the scoring system. We added the scores for the other preventive measures for a maximum value of four and a minimum value of zero. The goal of our exploratory model was to identify whether any group was more or less likely to meet guidelines overall, across different services lines.

Additional variables included were gender, race, and ethnicity from the registry form; participant's age, annual household income, highest level of education completed by the primary guardian(s), and type of health insurance from the parent report form; and current autism spectrum disorder (ASD) diagnosis, having hypersensitivity/overreaction to stimuli/emotionally reactive (referred to here on as ‘hypersensitivity’), whether or not hypersensitivity was a limiting problem, and intellectual function from the clinician form. On the clinician form, intellectual function was rated based on clinical judgement and other available information. Although testing results may have factored into the clinician's assessment, the rating was not based on testing alone. The intellectual function option ‘developmental delay’ was only used for children under six years of age, but children under six years were not restricted to ‘developmental delay’ and were described as having other levels of intellectual function as well. Hypersensitivity was based on clinical observation and parent report, and we collapsed the pair of hypersensitivity questions to identify an individual as with and affected, or without or unaffected, by hypersensitivity.

We restricted the sample to individuals under 21 years of age who were enrolled in the registry, and had a complete parent and clinician form. Participants who were missing responses to any of the included variables were removed, as were participants with invalid responses, or those whose responses contradicted other information entered in the form. For all outcome variables, we excluded those who answered “don't know” or “choose not to

answer." Finally, those who selected the invalid the invalid immunization response described above were excluded from the analytic sample.

We carried out our analyses out using SAS, version 9.4 © (English, Cary, NC). Due to the number of participants excluded, we ran a chi-squared or Fisher's exact test to identify significant differences between the samples. We provide both unadjusted bivariate and adjusted multiple regression analyses to compare preventive care outcomes by demographic characteristics. We tested for statistical significance using chi-squared or Fisher's exact test in the bivariate analysis, and AOR for the multiple regression analyses. A significance level of 0.05 was used for all significance tests.

We adjusted each of our multivariable regression models for all non-outcome variables. A Hosmer-Lemeshow goodness-of-fit test was calculated for each model; a significant deviation from the hypothesized values was not found in any of the models. We also examined conditional indexes for multicollinearity, all of which fell below 30. Our goal was to identify patterns important to preventive care, so we did not adjust for the number of tests run, as that approach could potentially mask a true significant association.

Results

Of the 898 individuals in the baseline longitudinal dataset, 592 had a registry form, parent form, and clinician form on file (Fig. 1). Of those, we excluded 69 21 years or older, and an additional 117 because they had missing or invalid data for key variables. Table 1 summarizes the differences between those with complete data (n = 406) and those with missing or invalid data (n = 117). The two groups differed significantly in the distributions of household income (P = 0.04), type of health insurance (P = 0.02), ASD (P = 0.04), and hypersensitivity (P = 0.01). The lack of an overall pattern of significant differences between the two groups suggests that the likelihood of a systematic bias in our selection of participants is small. None of our outcome variables differed significantly between the two groups.

Of the 406 included participants, three-quarters were male, over half of the sample was under 11 years of age, and there was a higher proportion of non-Hispanic white (74.6%) and Hispanic (13.3) participants than participants of other racial/ethnic groups (Table 1). Annual income was slightly skewed toward higher income brackets, indicating that our sample may have more resources to devote to their child's health care than people in the general population. Approximately two-thirds of primary guardians had a college or post-graduate degree. Most had private health insurance, and only 1.2% (n = 5) had no health insurance. These five participants were excluded from any further analysis due to their small sample size. Overall, 74.6% of the cohort received recommended dental care, 55.4% received recommended influenza vaccinations, 92.1% received recommended immunizations, and 24.4% engaged in sufficient PA, according to guidelines. Nearly all (98.0%) reported having a regular pediatrician or doctor. Most participants had two or three recommended preventive care outcomes, in addition to having a regular pediatrician or doctor.

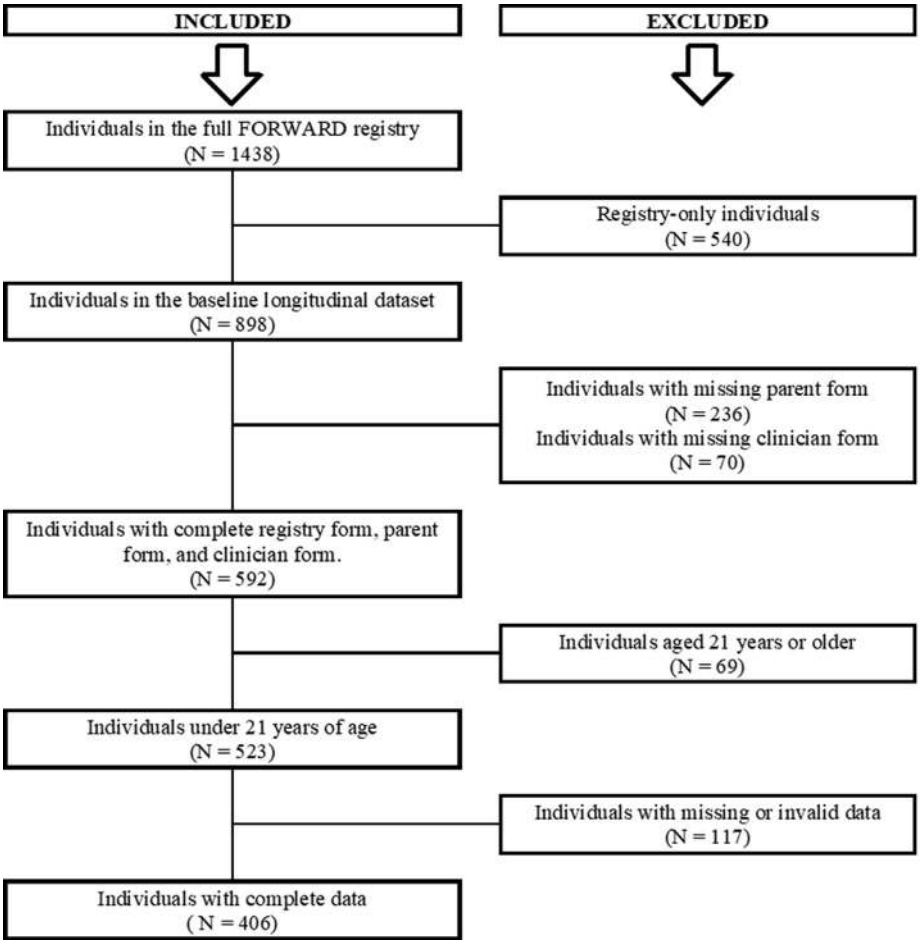

Fig. 1. Steps in the selection of individuals to be included in this study from the Fragile X Online Registry with Accessible Research Database, 2012–2015.

Table 1
Comparing baseline characteristics of included and excluded participants.

	Included Participants (n = 406)		Excluded Participants (n = 117)		
	n	%	n	%[c]	*P* value
Gender					0.93[d]
Male	310	76.4	89	76.1	
Female	96	23.7	27	23.1	
Age (years)					0.88[d]
0–5	123	30.3	33	28.2	
6–10	127	31.3	37	31.6	
11–15	98	24.1	32	27.4	
16–20	58	14.3	15	12.8	
Race/ethnicity					0.89[d]
White, non-Hispanic	303	74.6	85	72.7	
Black, non-Hispanic	20	4.9	5	4.3	
Hispanic, any race	54	13.3	16	13.7	
Other race, non-Hispanic	16	3.9	6	5.1	
Multi-racial, non-Hispanic	13	3.2	2	1.7	
Annual household income					**0.04**[d]
Under $25,000	34	8.4	10	8.6	
$25,000–$49,999	63	15.5	25	21.4	
$50,000–$74,999	69	17.0	23	19.7	
$75,000–$99,999	59	14.5	16	13.7	
$100,000–$149,999	73	18.0	8	6.8	
$150,000 or more	80	19.7	15	12.8	
Chose not to answer	28	6.9	12	10.3	
Highest level of education, primary guardian(s)					0.06[e]
High school or less	46	11.3	13	11.1	
Technical/some college	91	22.4	32	27.4	
College/Bachelor degree	133	32.8	35	29.9	
Post-graduate degree	136	33.5	24	20.5	
Don't know	0	0.0	1	0.9	
Type of health insurance					**0.02**[e]
Public	96	23.7	37	31.6	
Private	238	58.6	47	40.2	
Both	67	16.5	24	20.5	
None	5	1.2	0	0.0	
Last dental visit					0.86[e]
Within last year (met guidelines)	303	74.6	80	68.4	
Over 1 year, within 2	31	7.6	11	9.4	
Over 2 years, within 5	14	3.5	4	3.4	
Over 5 years	3	0.7	1	0.9	
Never	53	13.1	11	9.4	
Don't know	2	0.5	0	0.0	
Missing any immunization					0.37[e]
No (met guidelines)	374	92.1	101	8.6	
Yes, but plan to get missing ones	21	5.2	7	6.0	
No, and do not plan to get missing ones[f]	10	2.5	0	0.0	
Don't know	1	0.3	0	0.0	
Last influenza immunization					0.87[d]
Within last year (met guidelines)	225	55.4	56	47.9	
More than 1 year ago	84	20.7	24	20.5	
Never	89	21.9	26	22.2	
Don't know	8	2.0	3	2.6	
Physical activity in the past week (days)					0.72[d]
0	90	22.2	18	15.4	
1–2	123	30.3	34	29.1	
3–4	79	19.5	22	18.8	
5–7 (met guidelines)	99	24.4	28	23.9	
Don't know	15	3.7	6	5.1	
Has regular pediatrician/doctor					1.00[e]
Yes	398	98.0	105	89.7	
No	7	1.7	2	1.7	
Don't know	1	0.3	0	0.0	
Autism spectrum disorder diagnosis					**0.04**[d]
Yes	149	36.7	52	44.4	
No	225	55.4	44	37.6	
Don't know	32	7.9	8	6.8	
Intellectual function					0.16[e]
No intellectual disability	26	6.4	4	3.4	
Developmental delay, child under 6 only	65	16.0	10	8.6	
Borderline intellectual disability	32	7.9	9	7.7	
Mild intellectual disability	102	25.1	16	13.7	
Moderate intellectual disability	161	39.7	31	26.5	
Severe intellectual disability	18	4.4	10	8.6	
Profound intellectual disability	2	0.5	0	0.0	
Currently has hypersensitivity					**0.01**[d]

(*continued on next page*)

Table 1 (*continued*)

	Included Participants (n = 406)		Excluded Participants (n = 117)		
	n	%	n	%[c]	*P* value
Yes	288	70.9	80	68.4	
No	118	29.1	16	13.7	
Hypersensitivity is a limiting problem					0.48[d]
Yes	193	67.0	22	27.5	
No	95	33.0	8	10.0	
Total number of preventive care guidelines meta[a,g,b]					0.55[e]
0	1	0.4	1	1.4	
1	23	8.8	7	9.9	
2	90	34.4	29	40.9	
3	124	47.3	29	40.9	
4	24	9.2	5	7.0	

Boldface indicates statistical significance ($p < 0.05$).
[a] Includes dental care, immunization status, influenza vaccination, and physical activity.
[b] Participants who answered "don't know" or were missing dental, immunization, influenza vaccination, or physical activity were excluded, as were those to answered "No and do not plan to get missing ones" to the immunization question.
[c] Percent may not add to 100 due to missing values.
[d] Chi-square test.
[e] Fisher's exact test.
[f] Excluded from further analysis due to error on form.
[g] Children 6 years of age or older.

Fig. 2 shows unadjusted distributions of the demographic and health variables of those who had each preventive care outcome. The proportion of children and young adults receiving recommended dental care differed significantly across age (chi-squared test, P < 0.001), and was lower among children under six years of age with developmental delay (chi-squared test, P < 0.001), which may be an effect of age. Males were more likely to have received the influenza vaccine compared to females (chi-squared test, P = 0.03), and younger children were more likely to have received the influenza vaccine than older children (chi-squared test, P = 0.02). Participants with ASD were significantly less likely to have met PA guidelines (chi-squared test, P = 0.02) relative to participants without ASD.

The results for each of the multiple regression models examining dental care, immunization, influenza vaccination, and PA are shown in Table 2. After adjusting for all other variables, Model 1 found children under six years of age were less likely to have seen a dentist in the past year than children six to 10 years of age (AOR: 0.26; 95% CI: 0.11–0.60). Model 2 found that young adults 16–20 years of age were less likely than children six to 10 to have met immunization guidelines (AOR: 0.14; 95% CI: 0.02–0.96). Model 3 shows that non-Hispanic black participants (AOR: 0.25; 95% CI: 0.08–0.79) were less likely to have met influenza vaccination guidelines than non-Hispanic white participants. In Model 4, young adults 16–20 years of age (AOR: 0.29; 95% CI: 0.10–0.82), and those with ASD (AOR: 0.25; 95% CI: 0.10–0.59), were less likely to have met PA guidelines than children six to 10 years of age, and those without ASD. Individuals with and affected by hypersensitivity were also more likely to have met PA guidelines (AOR: 2.37; 95% CI: 1.15–4.88).

Our exploratory analysis (Model 5) found that young adults 16–20 years of age were less likely to have three or four preventive care measures compared to children six to 10 years of age (AOR: 0.34; 95% CI: 0.16–0.73). None of the other variables were significant when examined in our combined model. We observed wide CIs for several variables in the immunization model, and for 'other race' in the dental care model, as there was little variability between those variables and the outcome. Gender, household income, guardian education, health insurance status, and intellectual function were not significantly associated with meeting any preventive care guidelines in our regression models.

Discussion

Our study examined receipt of four preventive care outcomes; dental care, immunization status, influenza vaccination, and PA, all based on parent report. Table 3 compares our results to population-based studies examining these measures among children with special health care needs (CSHCN), children without special health care needs, and the general pediatric population. Approximately three-quarters of our clinic based cohort had seen a dentist within the past year, compared to 52–81% of all children who had seen a dentist at least once per year[25] and 45% of young adults with ID who had seen a dentist at least once per year.[11] Our results appear to contradict previously published results that dental care was the most common unmet health care need for CSHCN.[10] However, biannual visits are recommended as a preventive measure,[17] and while individuals in this cohort are visiting a dentist at least annually, they may not be attending every six months, or seeking preventive care services.

Like Houtrow et al.,[8] we found that well over 90% of parents reported that their child had a regular pediatrician or doctor, with our fragile X cohort falling slightly higher than both their estimation of CSHCN, and their estimation of all children. The likelihood of being fully immunized was high at 92.1%, which was much higher than that of children 19–35 months of age across the three other populations,[14,26] and higher than children 11–17 years of age with and without special health care needs.[6,13] The proportion of influenza vaccination in our cohort was similar to coverage estimations for the general pediatric population for the 2011–2012 influenza season.[27,28] Studies of PA have found that approximately 16% of all adolescents 12–17 years of age[29] and 12% of high school students[30] met 2008 PA guidelines. These estimates are much lower than the proportion meeting PA guidelines in our study, possibly due to the inexact match between the survey and preventive care guidelines.

Overall, we found that children and young adults with fragile X syndrome were as or more likely to meet common preventive care guidelines than typically developing children and young adults, which was unexpected given the previous research on CSHCN. However, the comparison groups included smaller proportions of non-Hispanic white and were less affluent than our sample, and our observations may be a result of our cohort having more resources and being drawn from specialty clinics, rather than being

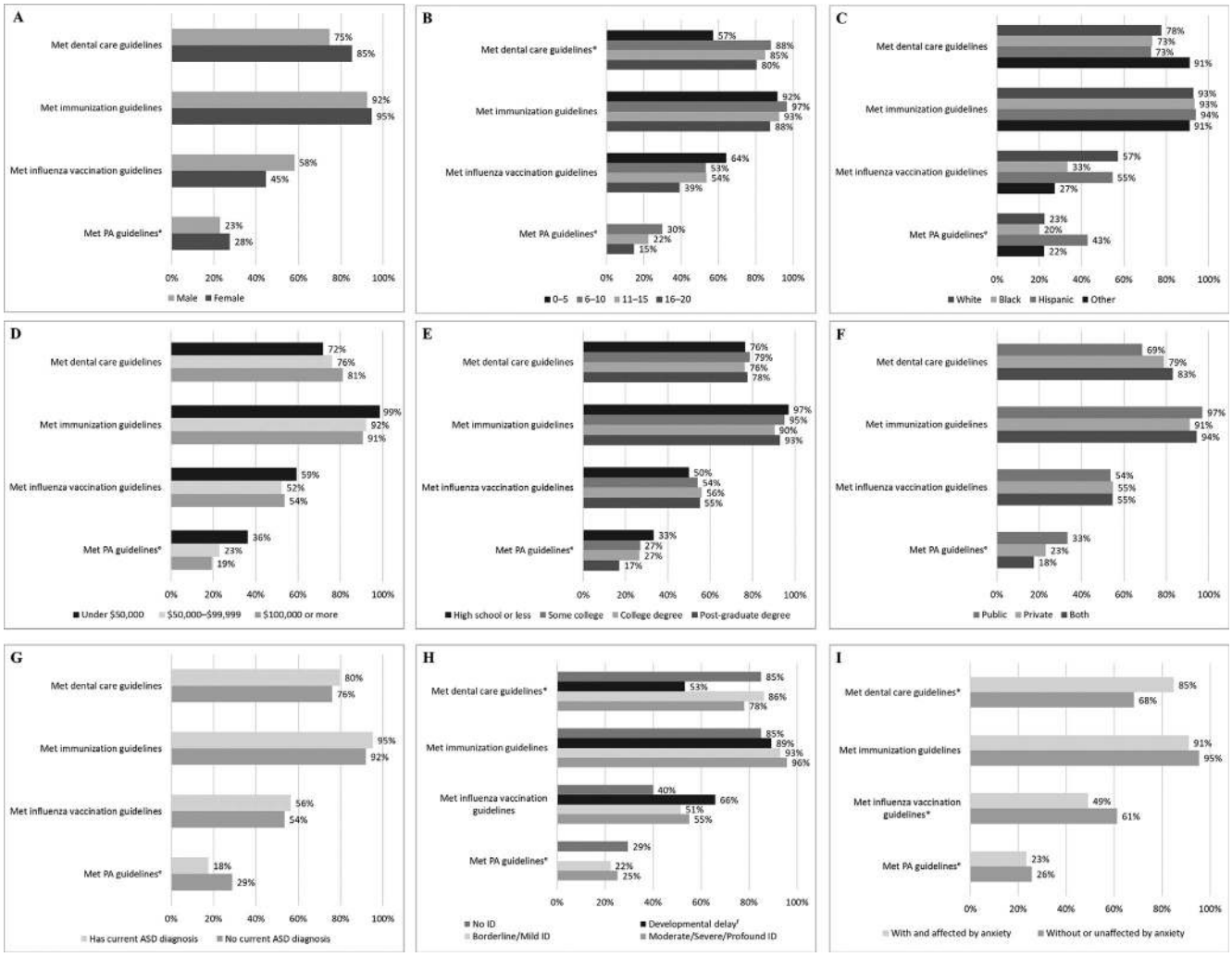

Fig. 2. Unadjusted distributions of participants who had each recommended preventive care outcome[a–d] by (A) gender, (B) age, (C) race/ethnicity, (D) income, (E) guardian education, (F) insurance, (G) autism spectrum disorder (ASD), (H) intellectual function, and (I) hypersensitivity; Fragile X Online Registry with Accessible Research Database (N = 316), 2012–2015.

[a]American Academy of Pediatrics Dentistry. Policy on the dental home. *Pediatr Dent*. 2012; 34(special issues):24–5.[17]

[b]Centers for Disease Control and Prevention. Recommended immunization schedule for children and adolescents aged 18 years or younger, United States, 2017. http://www.immunize.org/cdc/schedules/cdc-child-iz-schedule.html. Updated February 1, 2016. Accessed March 20, 2017.[18]

[c]Fiore AE, Uyeki TM, Broder K et al.; Advisory Committee on Immunization Practices. Prevention and control of influenza with vaccines: recommendations of the Advisory Committee on Immunization Practices (ACIP), 2010. *MMWR*. 2010; 59(RR-8):1–62.[19]

[d]U.S. Department of Health and Human Services. 2008 physical activity guidelines for Americans. https://health.gov/paguidelines/pdf/paguide.pdf. Accessed March 20, 2017.[20]

[e]Children 6 years of age or older.

[f]Children under 6 only.

* Chi-square test, $P < 0.05$.

Abbreviations: physical activity (PA); autism spectrum disorder (ASD); intellectual disability (ID).

unique to FXS.

We identified several possible disparities within the fragile X community. Younger children were less likely than children six to 10 years of age to have seen a dentist in the past year, which is consistent with disparities of dental care found in the general population.[24,31,32] Children six to 10 years of age were more likely than young adults 16–20 years of age to be engaged in sufficient PA; which has been documented in the general population.[33] Children six to 10 years of age were also more likely than young adults 16–20 years of age to have received recommended immunizations.

In our exploratory model combining different services, we found that children six to 10 years of age were more likely than young adults 16–20 to have three or four preventive care outcomes. The significantly lower odds of meeting guidelines among the late adolescent age group may reflect the difficulty many CSHCN face when transitioning from pediatric to adult care.[34] Such challenges include a lack of adult services, and services that are perceived to be inferior to pediatric care.[35] Our combined model is a unique feature of our analysis and we were unable to find additional literature to compare with our results. We believe this methodology provides valuable insight into groups who may not be meeting guidelines across the preventive care spectrum.

After adjusting for household income and guardian education, non-Hispanic black participants had lower influenza vaccination coverage than non-Hispanic white participants, while studies of the general population found similar or greater rates of influenza immunization among non-Hispanic black children compared to non-

Table 2
Factors associated with receiving recommended preventive care[d,e,f,g].

	Model 1: Met dental care guidelines	Model 2: Met immunization guidelines	Model 3: Met influenza vaccination guidelines	Model 4: Met PA guidelines[a]	Model 5: 0–2 vs 3 or 4 Preventive Measures[a]
	AOR[b] (95% CI)	AOR[b] (95% CI)	AOR[b] (95% CI)	AOR[b] (95% CI)	AOR[b] (95% CI)
Gender					
Male	Reference	Reference	Reference	Reference	Reference
Female	1.57 (0.73, 3.38)	2.22 (0.45, 11.02)	0.58 (0.31, 1.07)	0.97 (0.38, 2.47)	0.72 (0.34, 1.52)
Age (years)					
0–5	**0.26 (0.11, 0.60)**	0.17 (0.02, 1.32)	1.65 (0.78, 3.47)	Not applicable[a]	Not applicable[a]
6–10	Reference	Reference	Reference	Reference	Reference
11–15	0.8 (0.32, 1.97)	0.27 (0.03, 2.18)	0.97 (0.51, 1.87)	0.69 (0.31, 1.53)	0.61 (0.31, 1.20)
16–20	0.41 (0.17, 1.03)	**0.14 (0.02, 0.96)**	0.53 (0.25, 1.12)	**0.29 (0.10, 0.82)**	**0.34 (0.16, 0.73)**
Race/ethnicity					
White, non-Hispanic	Reference	Reference	Reference	Reference	Reference
Black, non-Hispanic	0.87 (0.26, 2.91)	0.59 (0.06, 5.92)	**0.25 (0.08, 0.79)**	0.45 (0.09, 2.42)	0.30 (0.08, 1.12)
Hispanic, any race	0.91 (0.40, 2.09)	2.09 (0.24, 18.38)	0.80 (0.39, 1.66)	2.66 (0.97, 7.26)	1.00 (0.38, 2.63)
Other race, non-Hispanic	2.73 (0.33, 22.76)	0.45 (0.05, 4.43)	0.38 (0.10, 1.36)	1.06 (0.22, 5.12)	0.38 (0.09, 1.56)
Multi-racial, non-Hispanic	0.74 (0.13, 4.02)	0.21 (0.02, 2.68)	1.59 (0.38, 6.65)	0.38 (0.04, 3.51)	0.79 (0.19, 3.27)
Annual household income					
$100,000 or more	Reference	Reference	Reference	Reference	Reference
$50,000–$99,999	1.12 (0.55, 2.29)	1.52 (0.42, 5.49)	0.94 (0.52, 1.69)	0.87 (0.36, 2.10)	1.52 (0.74, 3.13)
Under $50,000	0.85 (0.33, 2.19)	4.87 (0.55, 43.62)	1.70 (0.75, 3.87)	1.83 (0.59, 5.67)	0.96 (0.37, 2.48)
Highest level of education, primary guardian(s)					
Post-graduate degree	Reference	Reference	Reference	Reference	Reference
College degree	0.86 (0.41, 1.80)	0.32 (0.08, 1.24)	1.15 (0.62, 2.12)	1.30 (0.53, 3.21)	0.73 (0.36, 1.51)
Technical school/some college/associate's degree	1.02 (0.43, 2.46)	0.59 (0.09, 4.01)	0.84 (0.41, 1.72)	1.05 (0.36, 3.01)	0.95 (0.40, 2.24)
High school or less	1.02 (0.32, 3.29)	0.13 (0.01, 1.19)	0.80 (0.30, 2.15)	1.11 (0.30, 4.13)	1.92 (0.57, 6.42)
Health insurance					
Private	Reference	Reference	Reference	Reference	Reference
Both	1.91 (0.80, 4.57)	5.46 (0.60, 49.31)	0.66 (0.34, 1.29)	0.97 (0.35, 2.69)	1.35 (0.60, 3.00)
Public	0.87 (0.37, 2.01)	2.03 (0.29, 14.13)	0.60 (0.29, 1.26)	1.30 (0.49, 3.49)	0.98 (0.41, 2.32)
Autism spectrum disorder diagnosis					
No	Reference	Reference	Reference	Reference	Reference
Yes	1.20 (0.61, 2.34)	1.52 (0.34, 6.71)	1.00 (0.57, 1.75)	**0.25 (0.10, 0.59)**	0.67 (0.34, 1.33)
Intellectual function					
No intellectual disability	Reference	Reference	Reference	Reference	Reference
Developmental delay[c]	0.85 (0.21, 3.49)	3.81 (0.39, 36.93)	0.94 (0.26, 3.42)	Not applicable[¥]	Not applicable[c]
Borderline/mild intellectual disability	1.52 (0.44, 5.23)	6.76 (0.99, 46.29)	0.93 (0.34, 2.53)	0.64 (0.18, 2.27)	0.87 (0.28, 2.70)
Moderate/severe/profound intellectual disability	1.06 (0.29, 3.97)	5.04 (0.59, 43.24)	1.19 (0.40, 3.56)	1.09 (0.25, 4.74)	1.04 (0.29, 3.69)
With and affected by hypersensitivity					
No	Reference	Reference	Reference	Reference	Reference
Yes	0.98 (0.54, 1.78)	1.19 (0.37, 3.84)	0.85 (0.52, 1.41)	**2.37 (1.15, 4.88)**	0.89 (0.49, 1.64)

Boldface indicates statistical significance ($p < 0.05$).

[a] Children 6 years of age or older.

[b] OR adjusted for gender, age, race/ethnicity, annual household income, guardian level of education, type of health insurance, current autism spectrum disorder diagnosis, hypersensitivity status, and level of intellectual function.

[c] Children under 6 years of age only.

[d] American Academy of Pediatrics Dentistry. Policy on the dental home. *Pediatr Dent*. 2012; 34(special issues):24–5.[17].

[e] Centers for Disease Control and Prevention. Recommended immunization schedule for children and adolescents aged 18 years or younger, United States, 2017. http://www.immunize.org/cdc/schedules/cdc-child-iz-schedule.html. Updated February 1, 2016. Accessed March 20, 2017.[18].

[f] Fiore AE, Uyeki TM, Broder K et al.; Advisory Committee on Immunization Practices. Prevention and control of influenza with vaccines: recommendations of the Advisory Committee on Immunization Practices (ACIP), 2010. *MMWR*. 2010; 59(RR-8):1–62.[19].

[g] U.S. Department of Health and Human Services. 2008 physical activity guidelines for Americans. https://health.gov/paguidelines/pdf/paguide.pdf. Accessed March 20, 2017.[20].

Hispanic white children.[27,28] As expected, children and young adults with an ASD co-morbid diagnosis were less likely to meet the PA recommendations than their peers without as ASD diagnosis.[36] Conversely, children and young adults with or affected by hypersensitivity were more likely to have met PA recommendations than those without or unaffected by hypersensitivity. This contradicts our expectation that hypersensitivity would be a limiting problem for PA.

Despite children and young adults with FXS having similar or better rates of preventive care services and health behaviors compared to both CSHCN and the general pediatric population, our results show that continued effort on the part of health care providers is needed to increase compliance with preventive care guidelines. This is particularly important with regard to low PA, for which children with disabilities face a number of additional barriers, and low influenza vaccination, which falls well below *Healthy People 2020* goals. It is important to pay particular attention to children and young adults facing health disparities identified in our results, as well as adolescents in the vulnerable period of transition from pediatric to adult care.

Our study had several limitations. We used a clinic-based sample; parents who pursued a diagnosis for their child, attend specialty clinics, and answered survey questions may be more involved and have more resources than parents who do not attend clinic. All participants had health insurance, and our results may not reflect the experience of children living in lower income and

Table 3
Literature summary of recommended core health services and health behaviors[f,i,g,h].

Outcome	Children with Fragile X Syndrome	Children with Special Health Care Needs	Children without Special Health Care Needs	General Pediatric Population
Dental Care	74.6% of children under 21 years of age visited a dentist within the last year. (2012–2015)	45.1% of young adults 21–25 years of age visited a dentist at least once per year. (1997–2000)[e]	58.1% of young adults 21–25 years of age visited a dentist at least once per year. (1997–2000)[e]	51.9–81.1% of children 2–17 years of age visited a dentist or dental specialist in the past year. (2003)[j]
Immunization Status	92.1% of children under 21 years of age were not missing any immunizations.[k] (2012–2015)	12% of children 11–17 years of age received a tetanus booster, meningococcal, and human papillomavirus (HPV) vaccines. (2010–2012)[l,m] 59.0% of children 19–35 months of age received the 4:3:1:3:3:1[n] vaccination series. (2000–2002)[o]	12% of children 11–17 years of age received a tetanus booster, meningococcal, and HPV vaccines. (2010–2012)[l] 60.7% of children 19–35 months of age received the 4:3:1:3:3:1[n] vaccination series. (2000–2002)[o]	65.5% of children 19–35 months of age received the 4:3:1:3:3:1[n] vaccination series. (2002)[‡]
Influenza Vaccination	55.4% of children under 21 years of age received an influenza immunization within the past year. (2012–2015)			47.1–51.5% of children 6 months–17 years of age received an influenza vaccination. (2004–2012)[⸸⸸] 51.5–56.7% of children 6 months–17 years of age received an influenza vaccination. (2007–2012)[a]
Physical Activity	24.4% of children under 21 years of age exercised for at least 20 min 5–7 days in the past week. (2012–2015)			16.3% of all adolescents 12–17 years of age met 2008 physical activity guidelines. (1999–2006)[b] 12.2% of high school students met physical activity guidelines. (2010)[c]
Child has a regular pediatrician or doctor	98.0% of children under 21 years of age had a regular pediatrician or doctor. (2012–2015)	94.8% of children 3–17 years of age had a usual source of care. (2002–2003)[d]		89.6% of all children aged 3–17 years had a usual source of care. (2002–2003)[d]

[‡]Centers for Disease Control and Prevention. National, state, and urban area vaccination levels among children aged 19–35 months—United States, 2002. *MMWR*. 2003; 52(31):728–32.[26].
[⸸⸸]Santibanez TA, Lu PJ, O'Halloran A, Grabowsky M, Singleton JA. Trends in childhood influenza vaccination coverage—U.S., 2004–2012. *PHR*. 2014; 129:417–27.[27].
[a] Centers for Disease Control and Prevention. Surveillance of influenza vaccination coverage—United States, 2007–08 through 2011–12 influenza seasons. *MMWR*. 2013; 62(4):1–28.[28].
[b] Song M, Carroll DD, Fulton JE. Meeting the 2008 physical activity guidelines for Americans among U.S. youth. *Am J Prev Med*. 2013; 44(3):216–22.[29].
[c] Centers for Disease Control and Prevention. Physical activity levels of high school students—United States, 2010. *MMWR*. 2011; 60(23):773–7.[30].
[d] Houtrow AJ, Kim SE, Chen AY, Newacheck PW. Preventive health care for children with and without special health care needs. *Pediatrics*. 2007; 119(4):e821–8.[8].
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[f] American Academy of Pediatrics Dentistry. Policy on the dental home. *Pediatr Dent*. 2012; 34(special issues):24–5.[17].
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[h] U.S. Department of Health and Human Services. 2008 physical activity guidelines for Americans. https://health.gov/paguidelines/pdf/paguide.pdf. Accessed March 20, 2017.[20].
[i] Centers for Disease Control and Prevention. Recommended immunization schedule for children and adolescents aged 18 years or younger, United States, 2017. http://www.immunize.org/cdc/schedules/cdc-child-iz-schedule.html. Updated February 1, 2016. Accessed March 20, 2017.[18].
[j] Romaire MA, Bell JF, Huebner CE. Variations in children's dental service use based on four national health surveys. *Pediatrics*. 2012; 130(5):e1182–9.[25].
[k] The survey did not specify which immunizations were required to be considered fully immunized.
[l] McRee AL, Maslow, GR, Reiter PL. Receipt of recommended adolescent vaccines among youth with special health care needs. *Clin Pediatr (Phila)*. 2017; 56(5):451–60.[6].
[m] Reiter PL, McRee AL. Correlates of receiving recommended adolescent vaccines among youth with special health care needs: findings from a statewide survey. *Vaccine*. 2016; 34:3125–31.[13].
[n] 4:3:1:3:3:1vaccine series is composed of ≥4 doses of diphtheria and tetanus toxoids and pertussis vaccine, diphtheria and tetanus toxoids, and diphtheria and tetanus toxoids and acellular pertussis vaccine (DTP/DT/DTaP), ≥3 doses of poliovirus vaccine, ≥1 dose of measles-containing vaccine, ≥3 doses of *Haemophilus influenza* type b vaccine, ≥3 doses of hepatitis B vaccine, and ≥1 dose of varicella vaccine.
[o] O'Connor KS, Bramlett MD. Vaccination coverage by special health care needs status in young children. *Pediatrics*. 2008; 121:e768–74.[14].

lower education households. Our sample was limited to participants who answered all questions relevant to the analysis, excluding a large portion who differed significantly from the analytic sample on several characteristics. The outcome measures were based on parent report and were not otherwise validated, which can be unreliable, specifically regarding a child being fully immunized.[37] The FORWARD questionnaire also does not specify which immunizations are required to be considered fully immunized, nor does it collect information on dental visits or PA in a manner that is directly translatable to national guidelines. Finally, we identified a wording error on the form, which led us to exclude one of the response options for immunization status.

Conclusion

Our aim was to describe the current state of preventive care among children with fragile X syndrome, which has not been previously examined. We found that receipt of key preventive care services and practicing health behaviors among children and young adults with FXS appears to be similar to, or greater than children with special health care needs and the general population of children. However, the proportion meeting a number of these guidelines remains suboptimal, and there is uneven access based on demographic and health characteristics. Health providers may wish to use these findings to target gaps in care identified for each

services, and focus on educating parents on reaching recommended preventive care objectives. This may help improve health and reduce disparities for children with and without disabilities. Based on our results, further research into preventive care services using validated or direct measure of outcomes would be beneficial, as would research into barriers to preventive care and evaluating interventions specifically within this sub-population of children with special health care needs. Further analysis using our methodology of combined preventive care measures may also be useful in identifying vulnerable groups across services.

Disclaimer

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Declarations of interest

None.

Previous presentation

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City/State	Clinic
Phoenix, Arizona	Phoenix Children's Hospital
Long Beach, California	Long Beach Memorial Miller Children's and Women's Hospital Long Beach
Sacramento, California	UC Davis Health System
Stanford, California	Stanford University
Aurora, Colorado	Children's Hospital Colorado
Washington, DC	Children's National Medical Center
Miami, Florida	Mailman Center for Child Development
Atlanta, GA	Emory University School of Medicine
Chicago, Illinois	Rush University Medical Center
Iowa City, Iowa	University of Iowa Children's Hospital
Kansas City, Kansas	University of Kansas Medical Center
Louisville, Kentucky	Weisskopf Evaluation Center
Baltimore, Maryland	Kennedy Krieger Institute
Boston, Massachusetts	Boston Children's Hospital
Ann Arbor, Michigan	University of Michigan Hospitals and Clinics
Minneapolis, Minnesota	University of Minnesota
Staten Island, New York	New York State Institute for Basic Research in Developmental Disabilities
New York, NY	Seaver Autism Center for Research and Treatment at Mount Sinai
Durham, North Carolina	Duke University Medical Center
Cincinnati, Ohio	Cincinnati Fragile X Research and Treatment Center
Cleveland, Ohio	Cleveland Clinic Foundation
Elwyn, Pennsylvania	Elwyn Fragile X Center
Lewisburg, Pennsylvania	Geisinger Fragile X Center
Pittsburgh, Pennsylvania	Children's Hospital of Pittsburgh of UPMC
Greenville, SC	Greenwood Genetic Center – Greenville Office
Dallas, Texas	Children's Health and UT Southwestern Medical School

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