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Evaluation of Pediatrician Adherence to the American Academy of Pediatrics Health Supervision Guidelines for Down Syndrome

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Abstract

The American Academy of Pediatrics's guideline on health supervision for children with Down syndrome (DS) offers pediatricians guidance to improve detection of comorbid conditions. Pediatrician adherence has not yet been comprehensively evaluated. Medical records of 31 children with DS who received primary care at two urban academic clinic sites from 2008–2012 were reviewed. Data was extracted on adherence to age-specific individual guideline components for each subject by year-of-life (total 84 years-of-life). Overall adherence across all components was 83% (2001 guideline) and 67% (2011 guideline). Adherence to thyroid, hearing, vision, and developmental components was >85%, and anticipatory guidance regarding atlantoaxial instability and sexuality was <35%. Overall adherence was higher when a subject was younger and when a provider was an attending-level pediatrician.

Key Words: *Down syndrome; guidelines; adherence*

Down syndrome (DS) is the most common worldwide cause of intellectual disability and the most common chromosomal disorder affecting live-born infants, with an estimated live birth prevalence of 1 in 792 among children born in the United States (deGraaf, Buckley, & Skotko, 2015). Currently, there are around 89,000 people with DS under the age of 20 years living in the United States (deGraaf, Buckley, & Skotko, 2016), making it highly likely that most primary care pediatricians (PCP) will care for children with DS in their practice.

In addition to intellectual disability, children with DS may have comorbid and complex medical conditions. Hearing problems and otitis media, vision abnormalities and other eye disease, obstructive sleep apnea, and congenital heart disease are found in over half of all people with DS (Bull & the Committee on Genetics, 2011). Thyroid abnormalities, obesity, gastrointestinal atresia, celiac disease, leukemia, iron-deficiency anemia, seizures, atlantoaxial instability, and mood and behavioral disorders are also much more common compared to the general population (Bull & the Committee on Genetics, 2011). Advances in the surgical correction of congenital heart defects have resulted in a markedly increased survival rate in DS, almost doubling the lifespan with many individuals now living into their fifties (Hayes et al., 1997; Kucik et al., 2013). However, people with DS still have a shorter life expectancy when compared to adults with other causes of intellectual disability and adults in the general population (Yang, Rasmussen, & Friedman, 2002). Because many comorbid medical conditions commonly found in individuals with DS are not present at birth, persons with DS require timely and consistent health supervision throughout their lifespan. Optimal medical management is associated with improved quality of life and functioning among persons with DS (Bull & the Committee on Genetics, 2011; Roizen & Patterson, 2003).

With the aim of improving medical care for persons with DS, the American Academy of

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Pediatrics (AAP) developed guidelines for the appropriate delivery of health maintenance services for children with DS. These screening guidelines were first published in 2001 and were revised in 2011 (Bull & the Committee on Genetics, 2001; Bull & the Committee on Genetics, 2011) The guidelines emphasize the importance of monitoring, screening, and diagnosing medical conditions associated with DS that result in significant morbidity or mortality and also provide a plan for recommended health surveillance and anticipatory guidance (Bull & the Committee on Genetics, 2011). Despite the existence of these guidelines for over a decade, very little data evaluating their utilization in the primary care setting in the United States has been published. A prior study of children in Oklahoma and Nebraska with DS revealed poor (14%) adherence to thyroid screening (Fergeson et al., 2009). Santoro, Martin, Pleatman, and Hopkin (2016) examined general pediatricians' implementation of eight modified DS guideline components among 82 children with DS, revealing a statistically significant increase in adherence to five of the eight components after an educational intervention (Santoro et al., 2016). Skotko, Davidson, and Weintraub (2013) surveyed parents and reviewed prior records to determine the status of screening for five conditions by PCPs among children with DS under 3 years of age prior to presentation to a DS subspecialty clinic, finding that only 16.7% of patients were fully up-to-date. However, to our knowledge, no study has examined PCP records to determine the implementation of the full breadth of the AAP DS health supervision guidelines across all age groups among pediatricians in the United States.

The purpose of this study was to examine adherence to the AAP DS health supervision guidelines and to identify subject and clinician factors associated with guideline adherence.

Method

Research Design

We conducted a retrospective medical record review of children aged 0 through 17 years with DS who presented at either of two primary care clinics (Clinics A and B) associated with a major academic children's hospital in an urban setting from October 1, 2008, through September 30, 2012. All subjects seen at Clinic A were managed by pediatric residents with a supervisory attending onsite; Clinic B subjects were managed solely by primary care attending clinicians who specialize in the care of children with special health care needs (CSHCN). This study was approved by the hospital institutional review board.

Subjects

Subjects with DS receiving any care at either Clinic A or Clinic B in the study period were identified through the electronic medical records (EMR) system International Classification of Diseases, Ninth Revision, Clinical Modification (ICD-9) codes that indicated a diagnosis of Down syndrome (758.0, 758.0AG, 758.0AT, 758.0BG). Diagnoses of DS were confirmed by manual review.

Procedure

Data collection. Data obtained by EMR download included demographics, dates and type of visits, clinic site, distance from subject home to clinic, PCP level of experience (attending versus resident physician), problem lists, visit diagnoses, tests ordered, referrals placed, test results (including laboratory and radiology data), and dates of consultation notes and procedure/operative notes. Additionally, manual medical record review was used to identify missing information, review consultations and procedures, resolve discrepancies, and locate information not collected through the EMR download (such as assessment of symptoms, anticipatory guidance given, and documentation of care delivery at other locations). Each individual subject's EMR problem list was reviewed to identify all chronic medical comorbidities, and the total number of problems was recorded for each subject.

Guideline components. Based on the 2001 and 2011 consensus guidelines on DS health surveillance, we identified individual components of the AAP screening guidelines and the recommended ages for application of each component (Table 1). Because four components (annual hemoglobin measurement for all ages, annual atlantoaxial symptoms screening for all ages, annual celiac disease/gastrointestinal symptom screen for all ages, and screening polysomnogram prior to age 5) were added to the 2011 guideline with other modifications of the prior existing 2001 guideline, adherence was assessed with respect to both the 2001 and 2011 guideline sets. We defined adherence as being adequate if the record included written documentation, laboratory re-

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Table 1
American Academy of Pediatrics Down Syndrome Screening Guidelines for Children Aged 1 Year and Older

	2001	2011
Hematologic abnormalities	Hemoglobin annually from ≥13 years	Annually
Thyroid dysfunction	TSH annually	
Obstructive sleep apnea	Assess symptoms annually	Assess symptoms annually Screening polysomnogram by age 4 years
Atlantoaxial instability	Cervical x-ray at least once at 3–5 years	No screening x-ray Assess symptoms at each visit
Hearing problems	Hearing screen/referral: • Annually age 1–4 years • Once from age 5–12 years • Annually from age ≥13 years	Hearing screen/referral annually
Vision problems	Vision screen or referral: • Annually age 1–4 years • As needed age 5–12 years • Annually age ≥13 years	Visional screen or referral: • Annually age 1–5 years • Every 2 years from age 6–13 years • Every 3 years age from ≥14 years
Celiac disease or other GI problems	No recommendation	Assess symptoms annually Celiac screen NOT recommended for asymptomatic patients
Developmental problems	Early intervention referral by age 3 years Discuss transition to public school age 2–3 years	
Discussion about sexuality	Assess yearly if ≥12 years	

Note. TSH = thyroid stimulating hormone; GI = gastrointestinal.

sults, or radiologic reports pertaining to the guideline component (summarized in Table 2). Provider documentation of delivery of care of components at outside locations was also noted. Given that a PCP's role is to direct the medical home and oversee all care, we assigned the PCP "credit" for adequate screening if it was performed at any time during the study period, regardless of who the ordering provider may have been.

To evaluate adherence with guideline components, care delivery to each subject was examined over 1-year periods, henceforth "subject-years" (starting from the subject's closest birthday after 10/1/2008 to one day prior to the next birthday). Only full years within the data period were included in the subject-year review and each subject-year had to include a health supervision visit. Subject-years without a health supervision visit were not evaluated because of assumed inadequate opportunity for the PCP to fully address proper guideline implementation. Additionally, we identified no children with DS seen in these clinics who were <1 year of age during study planning, so we subsequently focused on the study guideline components relevant to children aged ≥1 year.

For each subject-year, we identified guideline components that should have been completed during that year based on subject age. Certain guideline components were not considered necessary components for that subject-year if (1) the subject had been previously diagnosed with the condition being screened for, (2) a test was ordered because the subject was symptomatic instead of for the purposes of screening, or (3) the screen was documented as reportedly completed at another site of care and with no results documentation available in our EMR.

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Table 2
Individual Guideline Component Criteria for Screening and Diagnosis

Guideline component	Definition of achieving screening criteria (any of the following)	Definition of meeting diagnostic criteria for given medical condition
Hematologic	• Documentation of laboratory data (hemoglobin) to screen for iron-deficiency anemia	• Documented anemias or other hematologic abnormalities • Prior CBC revealing abnormal hemoglobin level
Thyroid	• Orders placed for TSH • Documentation of laboratory data	• Documented hypothyroidism in medical record • Abnormal TSH results • Treatment with levothyroxine
Obstructive sleep apnea	• Discussion of sleep symptoms • Referral order for PSG [2011] • Completion of PSG [2011]	• Documented OSA in medical record • Prior documented PSG with OSA
Atlantoaxial instability	• Subjective screening for symptoms of AAI • Documented anticipatory guidance relevant to AAI • Neurologic exam specifically assessing for myelopathic signs • Cervical radiograph order or results [2001]	• Documented AAI or spinal abnormalities in the medical record • Abnormal radiographic results
Hearing	• Evidence of objective hearing screening (behavioral audiogram) • Referral for audiology assessment • Discussion of hearing assessment/capacity within medical record	• Documented hearing abnormalities in medical record • Abnormal results from formal audiological evaluation
Vision	• Evidence of objective vision screening • Referral to ophthalmology • Documented ophthalmological evaluation • Discussion of vision within medical record	• Documented vision abnormalities in medical record • Abnormal results from formal ophthalmologic evaluation
Gastrointestinal	• Discussion of any GI symptoms which might indicate dysphagia, celiac disease, etc. [2011]	• Documented GI abnormality • Abnormal celiac screening labs • Abnormal swallow study or other evaluation
Developmental	• Documentation of referral to and/or reception of ongoing services through early intervention • Documentation of discussion regarding transition to preschool and change from an individualized family service plan (IFSP) through early intervention to an individualized education plan (IEP) through the public school system	• Screening measure only, so no diagnosis formally tracked
Sexuality	• Discussion of sexuality and puberty with teenaged patients	• Screening measure only, so no diagnosis formally tracked

Note. CBC = complete blood count; TSH = thyroid stimulating hormone; OSA = obstructive sleep apnea; PSG = polysomnogram; AAI = atlantoaxial instability; GI = gastrointestinal.

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Table 3
Characteristics by Subject (n = *31)*

Characteristics	*n*	%
Sex		
Male	16	52
Female	15	48
Race/ethnicity		
Hispanic	20	65
Black, non-Hispanic	5	15
White, non-Hispanic	2	5
Other, non-Hispanic	4	13
Insurance		
Public	29	93
Private	2	7
Number of conditions on problem list		
1–2	7	23
3–5	8	26
5+	16	52

Associated factors. We examined the relationship of subject and clinician factors that might be potentially associated with guideline adherence. Subject demographic factors included sex, age group, race/ethnicity, and insurance status. We also attempted to consider the impact of potential markers of medical complexity such as number of conditions on the problem list, number of medical specialties seen, and number of primary care clinic visits. Finally, we assessed the role of clinic-related factors by looking at PCP level of experience (attending versus resident) and the distance of the clinic site from the subject's home (based on a rough calculation of miles from home zip code to clinic address).

Data analysis. All analyses were conducted using SAS 9.2 (Cary, NC). For each of the individual guideline components, an individual component adherence was calculated as the number of subject-years with appropriate component implementation divided by the total number of patient-years eligible for that component screening. For each subject-year, *overall adherence* (i.e., adherence across all components) was calculated as the number of completed guideline components within that patient-year divided by the total number of applicable guidelines within that patient-year. A *total adherence* was calculated for each guideline set as a whole (2001 and 2011) by averaging the *overall adherences* of all subject-years. Descriptive analyses, including frequency analysis and distribution analysis, were conducted for subjects' demographic characteristics, individual guideline components, potential predictors of the adherence, and number of comorbid conditions associated with DS.

T-tests were conducted to compare *overall adherence* by age group, sex, race/ethnicity, distance from home to clinic (<2 miles versus 3–10 miles), physician level of experience (attending versus resident level), and insurance (public versus private). Pearson correlation coefficients were calculated to examine the correlation between *overall adherence* and number of comorbid conditions, number of primary care visits, and number of specialists seen. A multivariate mixed model was used to examine the association between *overall adherence* to the 2001 guideline and potentially associated factors. Within-subject random effect was taken into account.

Results

Over the 4-year data collection period, 34 subjects with DS received care at these clinics and, among these, 31 had a health maintenance visit and thus were eligible for further evaluation. Most subjects (93%) had public health insurance and 65% were Hispanic (Table 3). Using records from these 31 subjects, we evaluated data encompassing 84 subject-years, including 25 subjects contributing 3 subject-years, 3 subjects with 2 subject-years, and 3 subjects with 1 subject-year. The majority of subject-years evaluated (58%) were for children aged 5–12 years (Table 4). Attending-level PCPs saw 57 subject-years (68%) at Clinic A during the study time-frame, whereas resident-level PCPs saw 27 subjects-years (32%) at Clinic B. Table 5 presents prevalence of comorbid conditions in our sample, including conditions identified during the screening processes under study (four cases of hypothyroidism and two cases of iron deficiency anemia).

Figure 1 displays adherence percentages for individual 2001 and 2011 guideline components. Individual component adherence was above 85% for thyroid, vision screening, and assessment of developmental service use. Guideline components with adherence below 50% included addressing sexuality in teenage subject-years (31%) and assessing for symptoms of atlantoaxial instability

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Table 4
Characteristics by Subject-Year (n = *84)*

Characteristics	*n*	%
Age Group		
1–4 years	20	24
5–12 years	49	58
≥13 years	15	18
Distance (home to clinic)		
0–2 miles	28	33
3–10 miles	56	67
Number of medical specialties seen		
0 specialties	16	19
1–2 specialties	34	40
3–4 specialties	27	32
≥5 specialties	7	8
Clinic		
A- Attending	57	68
B- Resident	27	32
Total number of primary care visits		
1 visit	22	26
2–3 visits	22	26
4–5 visits	22	26
≥6 visits	18	22

Table 5
Comorbid Conditions: Our Sample (n = *31) Compared to General Down Syndrome (DS) Population**

Condition	Our Sample (%)	General DS Population (%)
Hearing problems	42	75
Eye disease	68	60
Obstructive sleep apnea	42	50–79
Congenital heart disease	42	40–50
Otitis media	32	50–70
Thyroid disease	35**	4–18
Gastrointestinal atresia	13	12
Seizures	13	1–13
Iron-deficiency anemia	6***	10
Celiac disease	3	5
Atlantoaxial instability	13	1–2
Leukemia	0	1

*Prevalence data for comorbid conditions was quoted from the 2011 AAP guideline (Bull and the Committee on Genetics 2011). However, it should be noted that these numbers have been generated from healthcare data and may be inflated, as no populationwide surveys have looked at the true prevalence of these conditions among the "general" DS population.
**37% ($n = 4$) of cases were detected through screening during the study period.
***100% ($n = 2$) of cases were detected through screening during the study period.

(7%), which was a recommendation emphasized clearly in the 2011 guideline and a modification of the prior 2001 guideline that called for universal screening cervical x-rays for preschool-aged children. Of subjects seen in 2012, 4% still had screening cervical x-rays done even after dissolution of this 2001 recommendation. Additionally, even though broad celiac screening is not recommended in asymptomatic children with DS in either the 2001 or 2011 guidelines, celiac testing was nonetheless ordered for 19% of subjects (with no mention of concerning symptoms in provider notes).

Overall adherence to the 2001 guidelines was 83% (*SD* 19), compared to 67% (*SD* 20) for the 2011 guidelines. Higher *overall adherence* was found for children (subject-years) <13 years of age (1–4 years 86.5%; 5–12 years 88.7%) versus children (subject-years) ≥13 years of age (60.0%; t-test, $p < 0.0001$). Additionally, receipt of care from attending pediatricians was associated with significantly higher *overall adherence* to the 2001 guidelines (87.6%) when compared to residents (73.5%; t-test, $p = 0.0012$). Other postulated markers of disease severity (number of problems on problem list, number of primary care visits, and number of specialists seen during the subject-year) were not significantly correlated with *overall adherence* to the 2001 guideline (data not shown). Additionally, *overall adherence* to the 2001 guideline was not significantly different by subject sex or distance from home to clinic (data not shown).

A multivariate mixed model (Table 6) showed that age <13 years was associated with 23–29% higher guideline *overall adherence* compared to the adolescent age group (≥13 years; $p < 0.001$). Similarly, attending-level care was associated with 13% higher *overall adherence* to the 2001 guidelines when compared to resident-level care ($p < 0.006$). Sex, number of clinic visits, number of specialists seen, and distance from home to clinic were not

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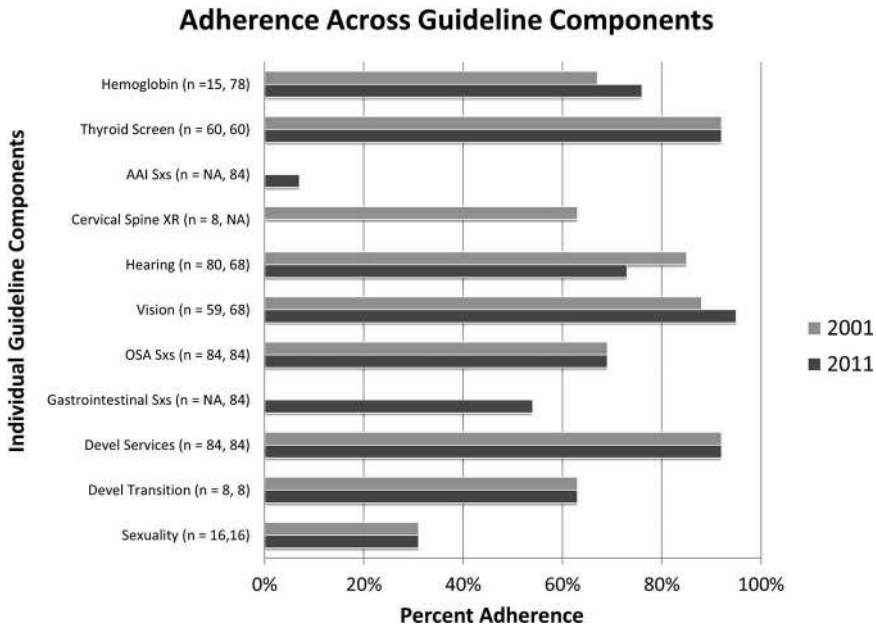

Figure 1. Individual guideline component adherence percentages. n = number of eligible subject-years for each component, with first number indicating 2001 data and second number indicating 2011 data; AAI = atlantoaxial instability; NA = not applicable; Sxs = symptoms; XR = x-ray; OSA = obstructive sleep apnea; Devel = developmental.

significantly associated with *overall adherence* (data not shown).

Discussion

This study presents a new method for evaluating adherence to DS guidelines. Although prior studies have evaluated select individual modified components over limited timeframes (Santoro et al., 2016), we assessed PCP adherence to each individual guideline component by subject-year over several years of a patient's life. As such, we were able to identify the individual screening components that were relevant to each particular subject during each year, with the goal of capturing a more precise measurement of guideline adherence among PCPs.

Table 6
Multivariate Mixed Model of 2001 Overall Guideline Adherence

	Effect Size		
Independent Variable	Estimate	(*SE*)	*p*-value
Age group			
1–4 years (vs. 13+ years)	22.8	(6.3)	<0.001
5–12 years (vs. 13+ years)	28.4	(5.2)	<0.001
Attending physician (vs. resident physician)	12.8	(4.4)	<0.006
Total primary care visits	0.03	(0.87)	0.82
Number of specialty types seen	0.6	(1.0)	0.58
Female (vs. male)	2.9	(3.7)	0.44
Home to clinic distance >2 miles	1.0	(4.4)	0.82
Intercept	53.3	(5.1)	<0.001

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In contrast to general trends of low adherence to AAP practice guidelines in other areas of pediatrics such as treating UTIs or screening for developmental issues at primary care visits (Cohen, Rivera, Davis, & Christakis, 2005; Sand et al., 2005), we observed that *overall adherence* to AAP DS guidelines was generally high (83% using 2001 AAP guideline). Additionally, our adherence findings are quite favorable in comparison to the international and adult literature on DS health surveillance. One potential explanation for high adherence in this instance might include the setting. This study was performed at a large academic medical center and the attending-level pediatricians involved in the study had a particular interest in caring for CSHCN. It is also therefore unsurprising that *overall adherence* among these attending-level PCPs was higher in comparison to resident-level physicians.

Our results that show experience-dependent screening adherence rates highlight two important points. First, even pediatricians who specialize in caring for CSHCN may be overlooking some age-appropriate screening guidelines when seeing subjects with DS, especially anticipatory guidance-based screening. However, it is important to point out that PCPs may have had conversations about these issues but did not document them fully, thus leading to seeming "failure" of adherence. Nonetheless, improvements in care could potentially focus on more routinely discussing issues like sexuality and screening for symptoms of gastrointestinal dysfunction and atlantoaxial instability, all of which had lower adherence rates (though the latter two were only initiated with release of the 2011 guideline). Second, residents need assistance implementing the AAP DS guideline. Santoro et al. (2016) showed that adherence rates are improved with the use of age-appropriate DS guideline checklists. Other simple interventions that could lead to improvements might also include the creation of a smart phone application to assist providers with screening or the use of templated notes within the EMR that provide prompts for age-appropriate screening. Further research should address reasons for poorer adherence, such as potential lack of awareness of the existence of guidelines, disinterest in or lack of full-time dedication to primary care among pediatric residents, resource-poor clinic settings, etc. Finally, education on DS comorbidities and recommended screening could be formally incorporated into the pediatric residency curriculum in a variety of ways, including online learning modules, in-person lectures, or regular dissemination of the 2011 AAP guidelines for Down syndrome and screening recommendations checklists to all residents in their continuity clinics at the beginning of each academic year.

The other factor that significantly impacted screening adherence rates was subject age. It is unclear exactly why older age was associated with significantly worse adherence, though the relatively poor adherence to sexuality-based anticipatory guidance may have skewed the teenaged sample toward worsened overall adherence. However, this finding is also consistent with data showing that typically developing adolescents are subject to lower preventative care guideline adherence by PCPs (Irwin, Adams, Park, & Naewacheck, 2009). Sexuality is a screening category that especially necessitates improvements given that youths with intellectual disability and other chronic conditions are more prone to becoming victims of sexual abuse (Chamberlain, Rauh, Passer, McGrath, & Burket, 1984; Surís, Resnick, Cassuto, & Blum, 1996). Very little information exists on the rates of sexual activity among adolescents with DS in particular, but a discussion with patients and their families is still called for at every adolescent health maintenance visit to address the issues of the potential for victimization, the need for safety during sexual activity, the prevention of sexually transmitted infections, and so forth.

We also found that factors that we speculated might complicate care (e.g., high numbers of comorbid conditions or specialists involved, living further from clinic, public insurance coverage as an indicator of lower socioeconomic status) did not contribute to lower adherence rates. However, the majority of subjects in this study were insured publically, without a high socioeconomic status comparison group. Previous research has shown that, when compared with other CSHCN, children with DS are more likely to have unmet health care needs and are less likely to have access to a medical home, and these differences were magnified further in instances of lower socioeconomic status and uninsured status (McGrath, Stransky, Cooley, & Moeschler, 2011). A future study with a more diverse population might be able to better determine if these factors impact adherence to health care guidelines in DS.

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When looking at guideline components individually, our results demonstrated that practitioners complied fairly well with vision and thyroid screening, both with over 85% adherence. Hearing screening also had high adherence rates, though this was one area where adherence fell when examining the 2011 guideline versus the 2001 guideline, likely because the 2011 guideline called for more frequent hearing screens than the earlier 2001 guideline. Weaknesses were seen with issues of anticipatory guidance, such as discussing sexuality and discussing symptoms of atlantoaxial instability, though it is possible such discussions did occur during the visit but were not documented. In comparison to the existing body of literature looking at use of health surveillance guidelines among persons with DS, this study reinforces the variability in implementation of particular individual recommendations, as others have reported. Studies of hearing-specific screening indicate that about two-thirds of children with DS in the United Kingdom are successfully screened (Stephen, Diskson, Kindley, Scott, & Charleton, 2007), but less than one-third of children with DS in Canada (Virji-Bubul, Eischmann, Kisly, Down, & Haslam, 2007) and less than one-third of adults with DS in the United States (Jensen, Taylor, & Davis, 2013) are screened appropriately. Thyroid screening rates among children with DS range from 13% in one United States study (Fergeson et al., 2009), to 71% in the United Kingdom (Varadkar, Bineham, & Lessing, 2003), to a pervasive lack of any screening in Israel (Wexler et al., 2009). When looking at adult DS thyroid screening rates, the numbers drop to $<$50% in the United Kingdom (Piachaud, Rohde, & Pasupathy, 1998) and Finland (Määttä et al., 2011) compared to a rate of 60% in the United States (Jensen et al., 2013). The United States study looking at primary care for adults with DS also highlighted particular weaknesses in screening for obstructive sleep apnea, atlantoaxial instability, and hearing/vision loss (Jensen et al., 2013).

Finally, in addition to simply assessing adherence rates, this study also emphasizes the utility of screening as evidenced by the many comorbid conditions identified during the relatively short study time frame. For example, among our 31 subjects, seven had pre-existing hypothyroidism but four more cases were detected among the remaining 24 subjects. Two cases of iron-deficiency anemia were also detected. Our review of records also revealed that some practitioners adopted screening patterns that were not dictated by the AAP health surveillance guidelines. For instance, some providers used laboratory tests to screen annually for leukemia and celiac disease (and, as specifically stated in clinic encounter notes, these patients had no symptoms), which is not recommended in the published guidelines (Bull & the Committee on Genetics, 2011).

Limitations

There were several limitations to this study. First, the patient population studied was seen by a very small number of physicians at only two urban clinic sites at an academic children's medical center; this limits the generalizability of findings. In particular, over two-thirds of subjects were seen by attending physicians who were especially skilled in the primary care of CSHCN. Additionally, the number of subjects with DS who were seen at these clinic sites during the study time frame was relatively low.

Second, although these AAP guidelines serve as an invaluable resource for pediatricians, they represent only a framework to build upon and should be individualized according to various patient needs. The individual guideline components were formulated based on existing research, but many are influenced heavily by consensus expert opinion and have not been studied extensively, making them subject to some interpretation. Importantly, it must be noted that a large-scale revision of the guidelines occurred during the midst of this study time period (2011), so expecting providers to have followed this later guideline is unreasonable and was analyzed only to better understand practice patterns. Thus, additional study is needed to understand adherence to the 2011 guidelines among community-based pediatricians on a larger scale.

A final limitation of this study was its reliance upon accurate documentation through an electronic medical records system. It is possible that guidelines may have been met, especially those that are anticipatory guidance-based, but not documented in clinician notes. If a subject received care at an outside institution during the study period, it is possible that some individual guideline components were met in actuality but not recorded in the EMR system. Nevertheless, we report high adherence with the guideline components that were evaluated.

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Directions for Future Research

More research is needed to identify factors contributing to inconsistent screening patterns and to test interventions that might improve adherence rates. This may range from continuing medical education on the topic, to the development of clinical tools to remind providers when certain guidelines should be implemented, to the empowerment of families to remind and educate primary care providers. In line with this last suggestion, a family-friendly AAP guideline checklist was recently developed that might serve as a reminder tool both for families and practitioners (Bull et al., 2016). Finally, greater utilization of multidisciplinary centers and subspecialty clinics that provide care to people with DS has also been shown to improve overall health surveillance for this population (Skotko, Davidson, & Weintraub 2013) though, unfortunately, disparities in access to subspecialists remains an issue for many people across the United States.

Conclusion

Despite the existence of the AAP's well-published guidelines for health surveillance in children with DS, only a handful of studies have examined adherence among primary care pediatricians in the United States. Our findings add to the growing body of literature in this area, but contrast with prior findings of fairly low adherence. Our study revealed mostly adequate adherence to screening components among pediatricians based out of a large academic medical center, but did identify several areas for potential improvement. Furthermore, our high *overall adherence* rate may be distorted based on the inclusion of a majority of pediatricians skilled in the care of CSHCN and a small number of subjects examined. This pilot sets the stage for a larger study across a range of primary care settings to provide even more insight into adherence rates to AAP DS guidelines in community primary care settings.

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