Preventive Services Received by Adolescents With Cystic Fibrosis and Sickle Cell Disease

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Objective: To determine the proportion of adolescents with cystic fibrosis (CF) or sickle cell disease (SCD) who reported speaking with their physicians about health-promoting and risky behaviors and whether the rate of discussions varied by whether the main physician was a primary care provider or specialist.

Hypothesis: Adolescents reporting a primary care provider as their main physician would be more likely to have received risk behavior counseling and other preventive services.

Design: Survey.

Setting: Comprehensive CF and SCD centers in 5 North Carolina referral hospitals.

Participants: Three hundred twenty-one (74%) of 437 adolescents aged 12 through 19 years (mean age, 15.6 years; 51% male) with CF or SCD identified through center registries.

Main Outcome Measures: Sources of health care, main physician, and recall of discussions with physicians regarding sexual issues, substance use, weight or dieting, safety issues, depression, and violence.

Results: Adolescents with CF (53%) or SCD (46%) most commonly reported a specialist as their main physician. For those (83%) who saw their main physician in the past year, adolescents with SCD reported counseling rates ranging from 43% for sexuality to 15% for weapon carrying or fighting. For adolescents with CF, rates ranged from 65% for weight and dieting to 30% for sexuality and 6% for weapon carrying or fighting. Adolescents whose main physician was a primary care provider were no more or less likely to report counseling for any topic (all \( P > .05 \)).

Conclusions: Physicians, regardless of specialty, infrequently discussed common behavioral issues with these adolescents with CF or SCD. A coordinated effort between primary care physicians and specialists may be helpful in delivering optimal preventive services to this population.


OFFICE-BASED preventive services, including screening and anticipatory guidance for risky behaviors, have been widely advocated as a means of reducing adverse health outcomes among adolescents and young adults. Previous studies of general adolescent populations have reported low rates of screening for risky behaviors. The low frequency of adolescent health care visits, especially preventive visits, has been cited as a reason for limited clinical preventive services, including risk behavior counseling.

Adolescents with chronic conditions are much more likely to interact with the health care system than are healthy adolescents, with an average of 8.8 visits per year compared with 2.2 for adolescents without a chronic condition. Greater visit frequency provides additional preventive opportunities for these adolescents. Preventive services for adolescents with chronic conditions are especially important because they are at higher risk for adverse health outcomes related to risky behavior. For example, cigarette smoking is associated with an accelerated decline in pulmonary function in persons with cystic fibrosis (CF) and an increased risk of acute chest syndrome in those with sickle cell disease (SCD). Cytotoxic medications used in a variety of conditions are associated with adverse fetal effects; thus, prevention of unplanned pregnancies is important.
SUBJECTS AND METHODS

STUDY POPULATION AND RECRUITMENT

The target population was all adolescents aged 12 through 19 years with CF (n=146) or SCD (n=292) identified through registries at all 5 major tertiary care centers in North Carolina. Families were introduced to the study by a mailed letter. Consent was obtained from the adolescent (and a parent or guardian if the adolescent was a minor) and data were collected during a clinic or hospital visit or by telephone for those without a visit during the 10-month study period (October 1994 to August 1995). Of 438 eligible adolescents, 35 (8%) declined to participate, 3 (1%) were excluded (2 lacked usable data forms and 1 had undergone bone marrow transplantation and no longer had SCD), and 79 (18%) could not be located, leaving 321 (74%) participants. Of these, 116 (36%) had CF and 205 (64%) had SCD, a 79% participation rate for those with CF and a 70% participation rate for those with SCD. Adolescents with CF were more likely to decline participation, and adolescents with SCD were more likely to be interviewed by telephone or not to be located. Nonparticipants were, on average, 2 months younger than participants. The sex distribution was similar (51% female vs 52% male). The study protocol was approved by the institutional review board at each of the 5 participating institutions.

INSTRUMENTS

Structured Interview

A structured interview was developed to determine sources of medical care, receipt of selected clinical preventive services (check-ups, sports physicals, immunizations, vision and hearing screens), and the adolescent's self-designated main physician. This instrument was reviewed both by clinicians familiar with the study population and by social scientists experienced in the design of survey instruments. It was then pilot-tested with an unrelated sample of adolescents with chronic conditions and revised for clarity. Formal reliability testing was not conducted. The term “main doctor” was chosen over the more common “usual source of care” or “usual provider” because it was the most clear to the adolescents in the pilot group. The question read: “Many people have a particular doctor or nurse that they consider their main or ‘own’ health provider. Do you have someone like that?”

Self-administered Questionnaire

Participants completed the Centers for Disease Control Youth Risk Behavior Survey, which assesses behaviors related to common adverse health outcomes in adolescents and adults. These include intentional and unintentional injury, tobacco use, alcohol and other drug use, sexual behaviors, physical inactivity, and diet. Summaries of the participants’ risk and protective behaviors are reported elsewhere. Additional questions were added to the survey to determine adolescent-reported physician counseling regarding each group of risk or protective behaviors. For example, at the end of the section on tobacco use, a question was added reading, “In the past 12 months, did your doctor talk with you about tobacco use?” Adolescents were instructed to refer to whichever physician they had reported as their “main doctor” during the preceding structured interview. Adolescents were not asked to distinguish whether they or the physician had initiated each conversation.

ADMINISTRATION

Adolescents were interviewed privately in the clinic. A research assistant unconnected with the clinic staff administered the structured interview and instructed the adolescent regarding completion of the questionnaire, then left the adolescent alone. Adolescents placed the completed questionnaire in a sealed envelope and then in a sealed box. An audio-recorded version of the questionnaire was also available for adolescents who requested it. Owing to the importance of assuring the adolescents’ anonymity, we made no attempt to correlate their self-reports with medical records or provider reports.

DATA ANALYSIS

The main outcomes were the adolescents' sources of care and receipt of anticipatory guidance or counseling for seat belt and helmet use; weight and dieting; lighting and weapon carrying; tobacco, alcohol, and other substance use; sexual behaviors; and suicide attempts and serious suicidal ideation. The counseling analyses included only those adolescents who reported seeing their main physician in the preceding year. Adolescents who indicated they were “unsure” if a counseling topic had been discussed were tabulated separately but then grouped with the positive responders for subsequent analyses and statistical testing. This was done to minimize false-negative responses. Because health care variables may be different for those with CF and SCD, all analyses were initially stratified according to disease category. Where no differences existed between them, the CF and SCD groups were combined in subsequent analyses. Preliminary analyses are reported as frequencies and proportions. Using 2-sided statistical tests, rates of counseling were then compared between those reporting a primary care provider (pediatrician, family physician, or nurse practitioner) or a specialist (pulmonologist or hematologist/oncologist) as their main physician. Odds ratios (ORs) and 95% confidence intervals (CIs) for counseling for comparisons between groups were calculated using Cornfield approximations. P<.05 was considered statistically significant. All data were analyzed using STATA 5.0 (STATA Corp, College Station, Tex).

pregnancy is essential. Nonetheless, studies in the 1970s and 1980s indicated that the primary health care needs of chronically ill youth, including risk behavior counseling and immunizations, were not receiving regular attention. A recent study of adult women with CF found that they were as likely as controls to be sexually active but were less likely to be using contraception. They also had limited knowledge of the effects of pregnancy on CF, and 33% of their pregnancies were reported as unplanned (a rate similar to that of the control group). Twenty-two percent had attempted to conceive at some time.
These earlier studies also reported that many adolescents with chronic conditions depended on their specialists at tertiary care centers for most or all of their ongoing health care needs. Some have suggested that, for patients with complex ongoing care needs such as those with chronic renal failure and SCD, the subspecialty center is the most appropriate place for most health care to be rendered. Others have suggested that primary care should best be received from a primary care physician in concert with the subspecialty team.

Because specialists may be more focused on disease-related issues than on general health promotion, we hypothesized that adolescents who reported that their main physician was a primary care provider would be more likely to have received preventive services, especially risk behavior screening and anticipatory guidance, than those who reported a specialist as their main physician. The objectives of our study were (1) to determine the usual source of health care for adolescents with CF or SCD, (2) to assess whether adolescents recalled speaking with their physicians about common adolescent health-promoting and risky behaviors, and (3) to determine whether receipt of preventive counseling varied by regular physician. We chose CF and SCD for this study because while they are both congenital conditions with usual onset of symptoms in early childhood, survival into young adulthood is increasingly common, making assessment of adolescent risk-taking increasingly important. Because both conditions have autosomal recessive inheritance, communication with adolescents regarding transmission risks and prevention of unplanned pregnancy is particularly important.

In addition to similar disease characteristics, the 2 conditions share similar health care delivery models. Multidisciplinary comprehensive care centers are prevalent and most patients are seen at a comprehensive center at least intermittently. Compared with national CF registry data, only 3% of North Carolina adolescents with CF were not included in the North Carolina care center records (S. C. FitzSimmons, PhD, written communication, 1995). Although there is no national SCD registry, it is estimated that about 80% of persons with SCD in North Carolina are known to one of the comprehensive SCD centers (J. Telfair, PhD, oral communication, 1995).

**RESULTS**

Participant demographic characteristics are summarized in Table 1. Adolescents with SCD were, on average, 0.7 years older than those with CF; the sex distribution was similar. As expected, there were large differences in the racial distribution of the groups. A specialist was the most common main or own physician for both those with CF (53%) and those with SCD (46%) (Table 2). Ten percent of those with CF and 23% with SCD reported they had no main or own physician. Among those with a main physician, 83% reported that they had seen that provider in the preceding year.

Thirty-one percent of those with CF and 15% with SCD reported having 1 or more other chronic conditions, most commonly asthma (reported by 8% with CF and 4% with SCD). In the preceding year, 43% of those with CF and 30% of those with SCD had sought care for health problems besides their CF or SCD. In addition, 31% of those with CF and 38% of those with SCD had been seen in the preceding year somewhere else besides the tertiary care center for their CF or SCD, as summarized in Table 2. When asked where they would go for a “minor health problem such as a sprained ankle or rash,” adolescents with CF would mostly commonly go to a pediatrician or family physician (56%), while adolescents with SCD would most likely go to the emergency department (33%) (Table 2).

Most patients reported receiving some form of preventive care. In the preceding year, 42% of those with CF and 51% of those with SCD reported a check-up or sports physical. Adolescents who reported having a main physician were no more likely to report having a pre-

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**Table 1. Participant Characteristics**

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>CF (n = 116)</th>
<th>SCD (n = 205)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean ± SD age, y</td>
<td>15.3 ± 2.1</td>
<td>15.8 ± 2.2</td>
</tr>
<tr>
<td>Sex, % female</td>
<td>51.3</td>
<td>48.3</td>
</tr>
<tr>
<td>Race, %</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Black</td>
<td>3.5</td>
<td>99.0</td>
</tr>
<tr>
<td>White</td>
<td>94.0</td>
<td>0.5</td>
</tr>
<tr>
<td>Other</td>
<td>2.6</td>
<td>0.5</td>
</tr>
<tr>
<td>Other chronic conditions, %</td>
<td>31.0</td>
<td>14.6</td>
</tr>
</tbody>
</table>

*CF indicates cystic fibrosis; SCD, sickle cell disease.*

**Table 2. Selected Health Care Services Utilization**

<table>
<thead>
<tr>
<th>Service</th>
<th>CF (n = 116)</th>
<th>SCD (n = 205)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Main physician</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Specialist</td>
<td>53</td>
<td>46</td>
</tr>
<tr>
<td>Family physician</td>
<td>10</td>
<td>9</td>
</tr>
<tr>
<td>Pediatrician</td>
<td>23</td>
<td>11</td>
</tr>
<tr>
<td>Nurse practitioner</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Other/unknown</td>
<td>5</td>
<td>10</td>
</tr>
<tr>
<td>None</td>
<td>10</td>
<td>23</td>
</tr>
<tr>
<td>Sought care for other health problems</td>
<td>43</td>
<td>30</td>
</tr>
<tr>
<td>Other sources of CF or SCD care besides CF or SCD center (could choose multiple)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pediatrician</td>
<td>47</td>
<td>27</td>
</tr>
<tr>
<td>Family physician</td>
<td>11</td>
<td>17</td>
</tr>
<tr>
<td>Nurse practitioner</td>
<td>0</td>
<td>3</td>
</tr>
<tr>
<td>Other (including other subspecialists)</td>
<td>31</td>
<td>16</td>
</tr>
<tr>
<td>Unknown</td>
<td>17</td>
<td>38</td>
</tr>
<tr>
<td>Where to go for minor problems</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pediatrician/family doctor</td>
<td>56</td>
<td>30</td>
</tr>
<tr>
<td>Emergency room</td>
<td>22</td>
<td>33</td>
</tr>
<tr>
<td>Another clinic</td>
<td>15</td>
<td>9</td>
</tr>
<tr>
<td>CF/SCD center</td>
<td>4</td>
<td>20</td>
</tr>
<tr>
<td>Other</td>
<td>3</td>
<td>8</td>
</tr>
<tr>
<td>Preventive care</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Check-up/sports physical (in last y)</td>
<td>42</td>
<td>51</td>
</tr>
<tr>
<td>≥1 immunization (in last 2 y)</td>
<td>86</td>
<td>51</td>
</tr>
</tbody>
</table>

*All data are presented as percentage. CF indicates cystic fibrosis; SCD, sickle cell disease.*
ventive visit than those without a main physician (OR=1.5; 95% CI, 0.9-2.6; P=.12). Influenza vaccine was received in the preceding 2 years by 79% of those with CF and 20% of those with SCD. One or more other immunizations in the preceding 2 years was reported by 32% of those with CF and 40% of those with SCD.

Adolescents reported that their regular providers infrequently addressed health-promoting or risky behaviors at an encounter occurring in the past year (Figure). Counting those who were unsure as positive responders, adolescents with SCD reported fairly consistent rates of counseling, ranging from a high of 43% for sexuality to a low of 15% for discussion of weapon carrying or fighting. Adolescents with CF reported more variable rates. Weight and dieting, a major concern in the routine care of CF, was reported to have been discussed with 65% of adolescents in the past year. Other issues were less commonly discussed, with a range from 30% for sexuality issues to 6% for weapon carrying or fighting. The median number of topics discussed was 2. No topics were discussed with 28% of adolescents, 1 topic with 23%, 2 topics with 15%, 3 topics with 10%, and 4 to 9 topics (of 9 possible) with 3% to 4% each.

Because providers may counsel differentially based on age or sex, we analyzed counseling rates stratified on these variables. For both CF and SCD, older age (15 years) was associated with higher rates of counseling for sexuality-related issues (47% vs 21%; OR=3.4; 95% CI, 2.0-5.8; P=.001). Males were, however, twice as likely to report having discussed tobacco use (32% vs 19%; 95% CI, 1.2-3.4; P=.007) and 1.7 times as likely to have discussed alcohol use (30% vs 20%; 95% CI, 1.0-2.9; P=.04) compared with females. The median number of topics discussed did not differ by disease category (CF or SCD), presence of other chronic health conditions, age, or sex.

Adolescents who reported that their main physician was a primary care provider (pediatrician, family physician, or nurse practitioner) were no more likely to report having received counseling for any behavior than adolescents who reported a specialist as their main physician (all P>.05). There was also no difference in the median number of topics discussed. Having had a check-up or sports physical was not associated with higher rates of counseling for any topic except for involvement with violence. Adolescents who had a check-up or sports physical were 2.5 times more likely to report having discussed this topic (18% vs 8%; 95% CI, 1.3-5.1; P=.008).

Overall rates of counseling, regardless of main physician, were low, below 50% for all topics, except for weight and dieting among those with CF. Contrary to our hypothesis, adolescents whose main physician was a primary care provider were no more likely to report counseling for any behavior than those who reported a specialist as their main physician. Those who reported a sports physical or check-up, visits for which prevention is a major goal, were no more likely to report counseling for any topic except violence. Based on these reports, communication between their main physicians and these adolescents with CF and SCD regarding common risk behaviors is not routinely occurring. This low level of counseling is consistent with previous reports from school-based studies, college students, and clinical practice medical record reviews.

In the general adolescent population, lack of preventive counseling has been attributed to time constraints, inadequate reimbursement, and physicians' lack of comfort and skills in adolescent care. Among those with CF and SCD, there may be additional barriers. Disease-related issues may predominate in both specialty and primary care visits. Some specialty centers may have provided more comprehensive care in the past, but new limits on visits imposed by managed care payers may have forced these centers to concentrate only on disease-related issues. Concurrently, because specialty centers provide multidisciplinary care, primary care providers may assume that preventive needs are being addressed there. For some topics, such as sexuality, where concerns regarding infertility or transmission of the condition to offspring may arise, primary care providers may wish to defer all counseling to the specialty center. Some providers may believe that adolescents with CF or SCD are more aware of their physical health and less likely to engage in health-compromising behaviors such as smoking.

Issues in the physician-patient relationship may also provide barriers to counseling regarding sensitive issues, especially at the specialty center. Because CF and SCD are lifelong conditions, many specialists have known the ado-
It is unlikely that large numbers of adolescents had been counseled in the more remote past. Many authorities, including the Guidelines for Adolescent Preventive Services1 and Bright Futures,2 recommend annual screening, since these behaviors often increase with increasing age. Third, counseling may have occurred but may not be recalled by the adolescent. We used a relatively short 1-year time frame to decrease recall problems. Adolescents who were unsure if counseling occurred were counted as having received counseling, so our estimates provide a “best-case” scenario for recalled counseling. Recalled counseling (rather than medical record documentation or provider report) may be the most appropriate measure of potentially useful counseling, because counseling that is not recalled is unlikely to have been effective in changing behavior. The relatively high rate of reported counseling regarding nutrition in CF (a concern related directly to CF) and the high rate of influenza immunizations suggest that the low numbers reported in the other categories are not primarily due to underreporting. Finally, the youngest patients in our sample were 12 years old when the study began. Although most authorities3-7 recommend risk behavior counseling for even preadolescents, some providers may not begin counseling until adolescents are older. Young participant age did not, however, seem to be a major factor contributing to lack of counseling in this study. Participants aged 15 years or older were no more likely to report counseling than younger participants, except for sexual behavior topics.

Although providing high levels of preventive services, including risk behavior counseling, has proved challenging,5-29 several strategies may be helpful for this population. In both the specialist and primary care setting, the American Medical Association’s Guidelines for Adolescent Preventive Services5 recommendations and implementation materials (including checklists and interview guides) can be used to screen adolescents more rapidly for a wide variety of issues and to provide guidance on managing areas of identified high risk. Clear discussions with parents and adolescents regarding confidentiality and routine time for adolescents alone with the physician may enhance adolescents’ willingness to share sensitive information.30-32 Physicians should examine and if necessary improve their skills and comfort level in counseling adolescents about sensitive issues such as sexuality and substance use. On a system level, programs such as the University of Minnesota’s “U-Special Kids,”33 which helps coordinate medical and other services for children and youth with complex chronic conditions, could improve communication between primary care physicians and specialty centers regarding counseling and other preventive services delivered and risks identified.

Provision of comprehensive care, including counseling and other preventive services, by specialty centers may become more difficult due to managed care constraints on visit frequency and services covered. Studies are underway to determine whether providing all care at a single site, such as a comprehensive SCD center, is associated with improved outcomes and lower cost than care distributed between community and specialty providers. Within these centers, nurse practitioners or physician assistants with special expertise in adolescent health care could provide counseling and other preventive ser-
Physicians can promote independence by assisting adolescents with gradually increasing responsibility for their health and health care decisions. Specialty centers that see significant numbers of adolescents could create regular adolescent-focused clinic sessions. Younger adolescents could “graduate” to these sessions for routine care. Designated adolescent clinics times signal to adolescents that the staff are sensitive to their needs and that it is appropriate to discuss adolescent issues. For parents and families, adolescent-focused clinics provide a gradual transition from parent-centered to adolescent-centered care.

For primary care physicians, it is important to remember that adolescents with chronic conditions like CF and SCD face the same developmental challenges and have the same preventive health care needs as adolescents without chronic conditions. They report risky behaviors at similar or slightly lower rates than those in the general adolescent population. They may also have additional concerns such as delayed maturation, difficulty in becoming independent from parents, or impaired fertility. Our results suggest that while these adolescents may receive some counseling from their specialty clinics, it is not universal and does not cover all important areas. A coordinated effort by both primary physicians and specialists may improve delivery of counseling and other preventive services to this population.

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