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The National Spina Bifida Patient Registry: Profile of a Large Cohort of Participants from the First 10 Clinics

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Objective To use data from the US National Spina Bifida Patient Registry (NSBPR) to describe variations in Contexts of Care, Processes of Care, and Health Outcomes among individuals with spina bifida (SB) receiving care in 10 clinics.

Study design Reported here are baseline cross-sectional data representing the first visit of 2172 participants from 10 specialized, multidisciplinary SB clinics participating in the NSBPR. We used descriptive statistics, the Fisher exact test, χ^2 test, and Wilcoxon rank-sum test to examine the data.

Results The mean age was 10.1 (SD 8.1) years with slightly more female subjects (52.5%). The majority was white (63.4%) and relied upon public insurance (53.5%). One-third had sacral lesions, 44.8% had mid-low lumbar lesions, and 24.9% had high lumbar and thoracic lesions. The most common surgery was ventricular shunt placement (65.7%). The most common bladder-management technique among those with bladder impairment was intermittent catheterization (69.0%). Almost 14% experienced a pressure ulcer in the last year. Of those ages 5 years or older with bowel or bladder impairments, almost 30% were continent of stool; a similar percentage was continent of urine. Most variables were associated with type of SB diagnosis.

Conclusion The NSBPR provides a cross section of a predominantly pediatric population of patients followed in specialized SB programs. There were wide variations in the variables studied and major differences in Context of Care, Processes of Care, and Health Outcomes by type of SB. Such wide variation and the differences by type of SB should be considered in future analyses of outcomes. *(J Pediatr 2015;166:444-50)*.

Spina bifida (SB) is one of the most common causes of disability among children and adolescents in the US and worldwide.[1] Approximately 1500 infants with SB are born yearly in the US,[2] reflecting an overall rate of 3.5 per 10 000[3] and a rate of 4.7, 3.2, and 2.6, respectively, in infants from Hispanic, non-Hispanic white, and non-Hispanic black mothers.[4] Myelomeningocele (MMC) accounts for the majority of individuals with SB identified at birth.[5] SB occurs in the early days of a pregnancy and results in a range of spinal cord and central nervous system impairments that lead to varying degrees of paralysis, limited mobility, impaired sensation, orthopedic problems (scoliosis, congenital hip dysplasia, and clubfeet), and bowel, bladder, and renal impairments.

Most children and adolescents with SB in industrialized countries currently survive, grow up, transition to adulthood, enter the adult health care system, and experience the typical health problems of adults.[6-8] However, data on their care and health status across the life span are limited. To overcome this limitation, and with the long-term goals of improving the health outcomes of people living with SB and building a foundation for ongoing research, the Centers for Disease Control and Prevention (CDC) funded 10 clinics to initiate the National Spina Bifida Patient Registry (NSBPR).[9] The feasibility of the project has now been established, with data collection operationalized at the first 10 clinics and 9 additional clinics.[9]

The purpose of this report is to describe baseline data of the participants enrolled in the registry from the 10 first clinics. A conceptual framework with 3 components was used for this study: Context of Care, Processes of Care, and Health Outcomes.[10] Context of Care included characteristics of the individual (demographic) and their chronic conditions (clinical). Processes of Care were defined as the actions that the individual, family, or health care provider implemented to affect clinical outcomes, such as types of surgeries, procedures, or bowel and bladder management techniques. Health Outcomes included

CDC	Centers for Disease Control and Prevention
MMC	Myelomeningocele
NSBPR	National Spina Bifida Patient Registry
SB	Spina bifida

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prevalence of pressure ulcers, bowel continence, and bladder continence. Understanding these factors and their interrelationships addresses the long-term goals of the NSBPR. The purpose of this study was to determine the distributions of Context of Care variables (demographic and clinical), Processes of Care (surgeries, bowel and bladder management techniques), and Health Outcomes (pressure ulcers, bowel and bladder continence) of individuals with SB in the NSBPR and to determine whether these variables differ by type of SB.

Methods

This descriptive study used baseline data obtained from the first NSBPR visit of the 2172 consented participants who were enrolled between March 2009 and June 2012 at the 10 first clinics. Patients were eligible for inclusion in the registry and in the current analysis if they had 1 of 4 SB types (MMC, meningocele, lipomyelomenigocele, or fatty filum) and received care from one of the participating clinics. The methods for the study, including those to monitor data quality and to obtain the institutional review/ethical board approval, have been reported in a previous publication.[9]

After consent was obtained, clinic personnel used a standardized tool to collect detailed data on 20 core registry questions, each with multiple data elements at all participating clinics.[9] The Initial Encounter Form completed only at the first NSBPR visit included basic demographic and diagnostic information as well as a lifetime history of selected common surgical procedures. The Annual Visit Form, completed at the time of the first and at each subsequent clinic visit, included clinical characteristics, anthropometrics, and insurance status. Data for the previous 12 months also were recorded on all surgeries performed (not just those listed on the initial form), on bowel- and bladder-management techniques currently used, and the targeted outcomes. The functional severity of the neurological impairment (level of lesion: sacral, low-lumbar, mid-lumbar, high-lumbar, and thoracic) was assessed by voluntary movement of hip, knee, or ankle. The analyses used in this study include data from both forms (Initial Encounter and Annual Visit Forms) collected at the first NSBPR visit.

Procedure and Analysis Strategies

Data from participants are collected by a combination of interview and medical record review. The data are entered into a custom-built, web-based SB electronic medical record at the participating clinics and transmitted to a central data center. At regular intervals, the data center, which hosts the web-based application, compiles, deidentifies, and transmits NSBPR data to the CDC for quality control and analysis.[9] Unless otherwise indicated, the full data set from the first 10 clinics was used for all analyses. To account for the effect of normal motor development, the mobility analysis was conducted using only participants aged 2 years and older. The variables "impaired bowel function" and "impaired bladder function" were created to identify those individuals whose SB affects their bowel and bladder functioning (Table I). Those individuals who were continent of stool and urine without using bowel or bladder management techniques were classified as "without impairments"; those with incontinence or who were using a listed technique to promote continence were classified as having "impaired bowel and/or impaired bladder function." Only data from those with impairments were used to describe the bowel- and bladder-management techniques. To account for development and expectations for socially acceptable continence, the sample of individuals with bowel and bladder impairments was further limited to patients ages 5 years and older for the continence analyses. For the second research question, SB type was collapsed into 2 groups: those with MMC and those with the non-MMC diagnoses (lipoMMC, meningocele, and fatty filum).

Data management and analyses were performed using SAS software 9.3 (SAS Institute, Cary, North Carolina).[11] Associations among categorical variables were examined by the Fisher exact test and χ^2 test, and continuous variables were compared by use of the Wilcoxon rank-sum test. Because of potential concerns regarding selection bias in the enrollment of patients into the NSBPR, data for individuals eligible but not enrolled in the NSBPR in 2012 ($n = 311$) were compared with data for individuals enrolled in the NSBPR and seen in 2012 ($n = 1602$). Data on eligible but not-enrolled individuals were only available for 2012, the first year that these data were collected. The 2 groups were compared by clinic for select demographic variables (age, sex, race, ethnicity, type of insurance), select clinical characteristics (diagnosis, level of lesion), and percent of eligible individuals enrolled. The analyses revealed that 2 clinics with the lowest rates of participation (63.2% and 69.2%) had more significant differences in demographic and clinical characteristics between eligible participants and nonparticipants (4 of 7). To determine whether selection bias influenced results for research question 2, analyses were conducted with and without these 2 clinics. Because there were no significant differences by type of SB in the total and reduced sample, the results of the total sample are reported. Although the sample used for the selection bias analysis did not exactly match the study sample for all first visits, it was the only comparison data available on eligible participants and nonparticipants.

Results

Context of Care

The distributions of demographic and clinical characteristics of the total sample of patients are reported in Table I. The mean age of the participants was 10.1 years (SD 8.1) and approximately 85% of them were younger than 18 years of age; the distribution of education levels corresponded with the age distribution. The largest cluster of school-aged participants was in grades 1-8. A majority of participants relied solely on public insurance. The balance had at least some private insurance. Almost 2 of 3 participants were non-Hispanic white, approximately 1 of 15 was non-