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Low vertebral ano-rectal cardiac tracheo-esophageal renal limb screening rates in children with anorectal malformations

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ABSTRACT

Background: The aim of this study was to establish the rate of screening for associated cardiac, vertebral, spinal cord, urologic, and limb anomalies vertebral ano-rectal cardiac tracheo-esophageal renal limb (VACTERL) in children with anorectal malformation (ARM).

Methods: We performed a retrospective cohort study using the Medicaid Analytic eXtract database which contains enrollment and utilization claims and demographic information from all Medicaid enrollees. Patients born between January 2005 and December 2008 with *International Classification of Diseases, Ninth Revision* codes for a diagnosis of ARM within 30 days of life, an ARM procedure code during the first year of life, and a minimum of 12 months of continuous enrollment were included. VACTERL screening was determined using *International Classification of Diseases, Ninth Revision* and Current Procedural Terminology codes for diagnostic tests used to detect these anomalies.

Results: A total of 406 patients were identified (231 males). Evaluation of the spinal vertebrae was performed in 94% of patients (381 of 406). Spinal cord evaluation was performed in 57% (231 of 406): 52% (121 of 231) received spinal ultrasound (US), 24% (56 of 231) received spinal magnetic resonance imaging, and 23% (54 of 231) received both. Sacral radiographs were performed in 8% (32 of 406) and 77% (313 of 406) underwent an echocardiogram. Genitourinary evaluation was performed in 84% (341 of 406): 67% (229 of 341) received renal US, 8% (27 of 341) received abdominal US, and 25% (85 of 341) received both. Limb evaluation was recorded in 19% (76 of 406). Multiple screening including an echocardiogram, spinal radiograph, spinal cord evaluation, and renal evaluation was performed in 45% (181 of 406); 2% (7 of 406) did not receive any screening tests.

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Conclusions: Low VACTERL screening in children with ARM suggests that associated anomalies may be undiagnosed which may lead to increased long-term morbidity.

Introduction

Anorectal malformations (ARMs) are rare congenital anomalies occurring in 1:4000-5000 live births.[1] ARMs are frequently associated with congenital anomalies affecting other organ systems. First described in 1973, vertebral ano-rectal cardiac tracheo-esophageal renal limb (VACTERL) association is an umbrella term for associated anomalies involving the vertebral column, anorectum, cardiac, tracheosophageal, renal system, and the limbs.[2-8] In children with ARM, the incidence of these associated anomalies is between 30% and 70% with more severe ARMs more likely to have associated anomalies.[9-13]

Children are classified as having the VACTERL association if they have three or more congenital anomalies. Although the VACTERL association is present in only 5%-31% of patients with an ARM, the acronym is a useful guide for initial evaluation, screening, and long-term follow-up in these patients to ensure all organs systems are evaluated.[10,14-16] In the immediate newborn period, these children may have anomalies which are immediately life threatening and will take priority in terms of management; however, without a mechanism to ensure that all organ systems are assessed in a systematic way, occult anomalies may be missed. Missed screening may lead to delayed diagnosis and intervention which can lead to increased morbidity and mortality and less informed and effective parental counseling.[17,18]

Screening rates for associated VACTERL anomalies in patients with ARM have not been reported in a broad population. The aim of this study was to investigate the rate of VACTERL screening in the newborn population with an ARM in the United States. Our hypothesis was that VACTERL screening rates in children with an ARM are low.

Materials and methods

We performed a retrospective cohort study using the 2005-2009 Medicaid Analytic eXtract (MAX), a healthcare utilization database that contains Medicaid enrollment and utilization claims and demographic information from all Medicaid enrollees from 50 states and the District of Columbia. For the present study, only the17 states with claims data usable for research were included (AK, HI, MT, NM, AZ, OR, WY, SD, IN, OK, AR, LA, MS, KY, NY, NH, and NJ).[19] The study cohort was derived from merging the MAX inpatient (IP) file, which contains IP admission records and the MAX other services file, which contains data on a variety of services, such as outpatient and clinic, hospice, home healthcare, laboratory and radiographic information, and physician and professional services. The Medicaid data set includes any procedure that generates a bill independent of which facility it is performed at. The *International Classification of Diseases, Ninth Revision* (ICD-9) diagnosis codes and Current Procedural Terminology codes were obtained from the merged data file.

Creation of the ARM cohort is described in Figure. Patients born between January 1, 2005 and December 31, 2008 with an ICD-9 diagnosis code for ARM documented within the first 30 days of life, a definitive ARM surgical procedure code within 1 year of life, and a minimum of 12 months of continuous enrollment in Medicaid were included (Table 1). The requirement for both the diagnosis and procedure code was used to improve the accuracy of identification of patients with ARM.[20] After establishing a final cohort of ARM patients, we investigated VACTERL screening (using a combination of Current Procedural Terminology and ICD-9 codes) to assess for the recommended screening tests for VACTERL associated anomalies after initial clinical evaluation (Table 2). Since many patients had the same radiologic investigation performed several times, the investigation and/or screening test with the earliest date was the test used in the final count. We investigated screening practices in the overall cohort, in males *versus* females, and in patients where a colostomy (ICD-9 codes 46.03, 46.10, 46.11, 46.13, 46.20, 46.21, and 46.23) was performed within the first week of life *versus* all other ARM patients. The colostomy subgroup analyses were performed because of the lack of diagnostic specificity of ICD-9 coding for ARMs. Since patients with a cloaca, rectobulbar, or rectoprostatic fistula are more likely to have a colostomy in the first seven days of life, compared with those with less complex anomalies (including perineal fistula or rectovestibular fistula),[21,22] a colostomy performed within the first seven days of life was used as a surrogate marker for increased severity of ARM.

Demographic characteristics and screening tests were described using frequencies and proportions. Pearson chi-square or Fisher's exact tests where appropriate, were used to compare screening between males and females, and patients with and without a colostomy in first week of life. A P value <0.05 was considered statistically significant. All analyses were performed using SAS 9.3 (Cary, NC). The institutional review board at our institution approved this study.

Results

Cohort development and demographics

Our initial search yielded 2794 patients with a diagnosis of ARM in the study period (Figure). A total of 2294 children did not have a definitive ARM procedure recorded and were therefore excluded. An additional 94 patients were excluded because of either noncontinuous enrollment ($n = 58$) or the definitive procedure not being performed in the first 12 months of life ($n = 36$). The final cohort included 406 patients of which 57% were male.