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Association for Academic Surgery

Lack of disparities in screening for associated anomalies in children with anorectal malformations

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ARTICLE INFO

Article history:
Received 1 December 2017
Received in revised form
18 April 2018
Accepted 4 May 2018
Available online xxx

Keywords:
Anorectal malformations
Congenital anomalies
VACTERL
Disparities
Neonatal

ABSTRACT

Introduction: Patients with anorectal malformations (ARM) often have associated congenital anomalies and should undergo several screening exams in the first year of life. We hypothesized that racial and socioeconomic disparities exist in the screening processes for these patients.

Methods: After IRB approval, a retrospective review of patients with ARM born between 2005 and 2016 was performed at a quaternary care children's hospital. Demographics including gender, race, insurance, and zip code were collected. Zip code was used as a surrogate for median income. Chart review was performed to identify anomaly type and whether Vertebral defects, Anorectal malformations, Cardiac defects, Tracheo-Esophageal fistula, Renal anomalies, and Limb abnormalities screening was performed within 1 y of age. Descriptive statistics and chi square analyses were performed.

Results: One hundred patients (59% male, 68% low malformation) were identified. African American and Caucasian subjects represented 41% and 40% of the population, respectively. Overall, 68 of 100 patients had at least one screening test for each of the Vertebral defects, Anorectal malformations, Cardiac defects, Tracheo-Esophageal fistula, Renal anomalies, and Limb abnormalities associations.

Although some minor differences were noted (more African Americans received skeletal survey than Caucasians, 80.5% *versus* 60%, $P = 0.00335$), no pattern of systematic bias in the receipt or timing of screening was evident based on race, insurance, or income.

Conclusions: There do not appear to be racial or socioeconomic disparities in screening for associated anomalies in patients with ARM. However, overall gaps in screening still exist, and work must be carried out to appropriately screen all patients for associated anomalies.

Presented at the 13th Annual Academic Surgical Congress, Jacksonville, FL, January 2018.

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0022-4804/$ – see front matter

https://doi.org/10.1016/j.jss.2018.05.008

Introduction

Anorectal malformations (ARM) are a group of rare congenital defects that result from abnormal development of the hindgut, allantois, and Mullerian ducts. ARM have been reported to occur in approximately one of every 4000-5000 live births, with a slight male predominance.[1] Although ARM represent a rare category of congenital defect, they are frequently associated with other defects. The Vertebral defects, Anorectal malformations, Cardiac defects, Tracheo-Esophageal fistula, Renal anomalies, and Limb abnormalities (VACTERL) association was first described in 1973 as the nonrandom association of vertebral, anorectal, cardiac, tracheoesophageal fistula/esophageal atresia (TEF/EA), renal, and limb anomalies.[2] Patients are classified as exhibiting the VACTERL association if they are diagnosed with three or more of these associated anomalies. The rate of VACTERL-associated anomalies in patients with ARM has been reported as high as 71%.[3-6] High lesions have been demonstrated to have a higher incidence of associated anomalies compared with low lesions.[6]

The presence of undiagnosed anomalies in patients with ARM is a potential source of morbidity and mortality that extends well into adulthood, emphasizing the importance of screening protocols.[7,8] Several screening recommendations have been suggested, including screening for other anomalies in patients with two VACTERL anomalies, screening all patients with TEF/EA and ARM, and screening all ARM for tethered cord.[9,10] Implementation of screening protocols has been demonstrated to improve anomaly detection in patients with ARM, although questions still remain about ideal screening modality and timing.[11]

Socioeconomic status (SES) is recognized as an important determinant of pediatric health outcomes. Race and income impact both health status and utilization of health services, with lower SES children less frequently using health resources.[12,13] Lower SES has been associated with worse outcomes in children in a wide variety of health parameters.[14] Previous studies have demonstrated that children with public insurance were less likely to receive specialty care than those with private insurance.[15-18] We hypothesized that lower SES ARM patients received less VACTERL screening.

Materials and methods

The University of Tennessee Health Science Center Institutional Review Board approved this study and, in accordance with the Code of Federal Regulations Title 45 (part 46, subpart D), granted waiver of informed consent. A single-institutional, retrospective cohort study was performed for patients born between 2005 and 2016 at an urban, quaternary care, freestanding children's hospital. Patients with ARM were identified using International Classification of Diseases 9 code 751.2 and International Classification of Diseases 10 codes Q42.3 and Q43.6. Diagnoses were then confirmed by individual electronic medical record review. Patients were excluded if the diagnosis was not documented in the medical record, if surgery was performed before 2005 (when the electronic medical record was implemented), or if there was no documentation of screening studies in patients who were transferred from other institutions. Patients with cloaca were also excluded. Demographics including gender, race, insurance, and zip code were extracted from the medical record. Zip code of residence was used as a surrogate for median income based on currently available census data.[19]

Chart review was performed to specify ARM anomaly type and to identify receipt and timing of VACTERL screening studies, including chest X-ray, skeletal survey, "babygram", renal ultrasound, echocardiogram, spine ultrasound, and spine magnetic resonance imaging (MRI). We used skeletal survey and babygram for vertebral anomaly screening, chest X-ray for TEF screening, echocardiogram for cardiac screening, renal ultrasound for renal anomalies, skeletal survey for limb anomalies, and spine ultrasound and MRI for spinal anomaly screening. ARM were grouped into "low" or "high" using modifications of the Wingspread and Krickenbeck classification systems. Low malformations include anal stenosis and perineal and vestibular fistula. High malformations include rectourethral and rectovesicular fistulas.

Demographics and screening tests are reported with descriptive statistics and chi square analyses were performed. Nonparametric tests were used where appropriate. A P-value <0.05 was considered statistically significant. All analyses were conducted in SAS 9.3 (Cary, NC).

Table 1 – Demographics.

Gender	% ($n = 100$)
Female	41%
Male	59%
Race	
African American	41%
Caucasian	40%
Other	18%
Low anomaly	68%
Anal stenosis	3%
Perineal fistula	45%
Vestibular fistula	20%
High anomaly	24%
Rectourethral fistula	13%
Rectovesicular fistula	3%
Rectovaginal fistula	2%
No fistula	6%
Other/unknown anomaly	8%
Insurance	
Government	67%
Private pay	12%
Commercial	18%
Missing	3%
Median income	
<25k	2%
25k-35k	33%
35k-50k	41%
>50k	19%