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Prevalence and ethnic variation of pre-auricular sinuses in children

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ABSTRACT

Objectives: Pre-auricular sinus (PAS) describes a congenital ear malformation presenting as a pit or sinus that may become infected, often requiring antibiotics and/or surgical excision. Although the presentation of PAS in otolaryngology clinics is not uncommon, there is limited epidemiological data regarding this malformation in children. Some evidence also suggests a potential ethnic variation in prevalence rates and potential heritability patterns within families, however these have yet to be proven. This study is the first to use pediatric population level data to investigate prevalence rate, ethnic variation, and to explore the unproven hypotheses of a genetic basis of PAS.

Methods: In this prospective cross-sectional study, we enrolled 1106 subjects (mean age = 6.8, male-to-female ratio = 1.15:1) between June and September 2014. Subjects were recruited from B.C. Children's Hospital in Vancouver, Canada. Inclusion criteria was children <18 years of age; exclusion criteria was those seeking care for PAS. Subjects were visually inspected for the presence of PAS by clinical observers followed by verbal questionnaire (demographics, self-identified ethnicity, family history of PAS, chronic medical conditions). Data analysis utilized Pearson Chi-Square Test to determine the potential ethnic variation, and odds ratios of family history were used to determine a potential genetic basis.

Results: 26 (7 bilateral, 19 unilateral) of 1106 subjects were positive for PAS (2.4%). Using Pearson Chi-Square Test, a significant ethnic variation was found to exist (χ^2 (6,N = 1106) = 22.80, $p < 0.0001$), with Asians having the highest prevalence (6.6%), followed by African Americans (4.5%), Middle Easterners (3.4%), First Nations (2.0%), and Caucasians (1.2%). None were found in South Asians (n = 124) or Latin Americans (n = 18). Subjects with positive family history of PAS had greater likelihood of having PAS (OR = 16.7, 95% CI = 7.3–38.5, $p < 0.0001$). There was also stronger association between family history and bilateral PAS (OR = 26.5, 95% CI = 5.8–121.7, $p < 0.0001$) compared to unilateral PAS (OR = 12.2, 95% CI = 4.6–32.5, $p < 0.0001$).

Conclusions: This was the largest pediatric population level study to date, and showed the prevalence of PAS was 2.4% in this pediatric population, whose ethnic diversity was representative of B.C.'s community. A significant ethnic variation existed and associations between family history and PAS suggested a potential genetic basis, particularly with bilateral PAS.

1. Introduction

1.1. Background

The pre-auricular sinus (PAS) is a congenital malformation of the ear typically characterized by a small pit or sinus opening adjacent to the external ear on the anterior margin of the ascending limb of the helix, which may lead deeply to an epithelial-lined cyst [1,2]. This congenital abnormality is believed to result from improper fusion of the first and second branchial arch derivatives (Hillocks of His) during the sixth week of gestation [2]. There are less common variants described, including an opening on the posterior margin of the helix, lobe, tragus, or superior to the crus [2–4]. While many patients are asymptomatic, a PAS is susceptible to infection by gram-positive bacteria, with patients often presenting with symptoms of sinus discharge, swelling, and pain [2]. Typically those patients with an infected PAS require antibiotics, drainage, and/or complete surgical excision to reduce the risk of infection recurrence, estimated at up to 42% [5,6]. PAS may also be associated with other ear or kidney malformations and syndromes, the most notable being Branchio-Oto-Renal (BOR) syndrome or Trisomy 22 [5,7]. PAS may present as a unilateral or bilateral abnormality.

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1.2. Literature review

The prevalence of isolated PAS in the pediatric population remains unclear, as this has never been specifically investigated. The overall prevalence in the United States is estimated to be 0.1 to 0.9% [2]. Worldwide, there appears to be a potential ethnic variation in prevalence rates suggested by the higher reported prevalence rates in Africa (4–10%), Taiwan (1.6–2.5%), and Malaysia (1.1%) compared to Hungary and England (0.47% to 0.9%) [2,8–12]. In a Malaysian study of soldiers, an ethnic variation between Chinese, Malay, and Indian adult males was described, further supporting the hypothesis of ethnic variability in prevalence [12].

There exists limited data supporting a potential genetic basis for PAS. These congenital abnormalities are believed to occur spontaneously or through inheritance and likely follow an incomplete autosomal dominant pattern with reduced penetrance in hereditary cases [2,5]. Bilateral PAS is estimated to occur in 25–50% of cases, and is thought to be more likely hereditary than unilateral PAS, which is presumed to be more commonly spontaneous and non-hereditary [2,5].

1.3. Study objectives

The purpose inquiries of this study were to: (1) estimate the prevalence of isolated PAS in the pediatric population, (2) to determine whether or not an ethnic variation in prevalence rates exists, and (3) to obtain population-level data to further support the hypothesized but currently unproven genetic basis for this malformation in children. The ethnic diversity of the pediatric population served by B.C. Children's Hospital in Vancouver provided a unique opportunity to explore this condition.

2. Methods

2.1. Subject recruitment

This cross-sectional observational study took place in the highest-traffic pedestrian areas in B.C. Children's Hospital from June to September 2014. Subjects were recruited from the waiting areas of the main hospital entranceway, Emergency Department, Otolaryngology clinic, and Orthopedics clinic. These areas were selected for their high patient turnover rate. Subjects were recruited in systematic fashion with two observers approaching every child in the waiting rooms of each department before moving to the next. The inclusion criteria for the study was all children under the age of 18 at the time of screening, regardless of clinical disorder or reason for hospital visit (seeking medical care, visiting inpatients, or waiting with the child needing care). The exclusion criteria for the study was subjects whose purpose of visit was for care specifically related to a known PAS. The study had full approval from the institutional Research and Ethics Committee.

2.2. Data collection

Carers of potential subjects were given an explanation of the study and asked to give verbal consent for their child's participation. At the time of consent, enrolled subjects were visually inspected by two clinical observers for the presence of a unilateral or bilateral PAS. Inspection was followed by a verbal questionnaire answered by the carer (see Table 1). Questions pertained to subject demographics, family history of PAS (known presence in a first-degree relative), pre-existing medical conditions, and self-identified ethnicity. Subjects identified as positive for PAS were asked additional questions including history of prior infection, their awareness of having a PAS, and any known hearing deficit.

2.3. Data analysis

Based on their response for self-identified ethnicity, subjects were grouped into one of eight categories based on Census Canada descriptors: African American, Asian, Caucasian, First Nations, Latin American, Middle Eastern, Mixed, and South Asian (see Table 2) [16]. Mixed subjects were defined as being part of multiple ethnic groups.

Data was collected verbally and recorded electronically using Microsoft Excel. Statistical analysis was performed using commercially available computer software package (SPSS for Windows version 22.0, SPSS Inc., Chicago, Illinois). To evaluate for an ethnic variation in prevalence rates, cross-tabulation using Pearson Chi-Square Test was used (mixed populations were excluded from this test). To evaluate for a potential genetic basis, family history, expressed through odds ratios, was used. $p < 0.05$ was considered statistically significant.

3. Results

3.1. Recruitment

1106 of 1111 recruited subjects were included in the study (male to female ratio, 1.15:1; mean age, 6.8 years [range, 0.01–17 years]). There were five withdrawals, defined as subjects enrolled but unable to complete the visual inspection and/or the age, gender, and self-identified ethnicity questions of the survey. There were 13 exclusions: four subjects were seeking care for an infected PAS and nine were 18 years of age or older. There were 94 refusals to take part in the study in which carers of potential subjects refused to give consent for personal reasons, commonly because of a perceived lack of time to participate or lack of interest in the study question. Table 3 highlights the population characteristics compared to the overall British Columbia population from which the patients were recruited, as well as comparisons to the US population [13,16–18].

3.2. Prevalence rates and ethnic variation

26 of the 1106 subjects included in the study were positive for a PAS (17 male, 9 female), demonstrating an overall prevalence of 2.4%. 7 were bilateral and 19 were unilateral (8 right-sided, 11 left-sided). 1 of the 19 unilateral subjects had a right-sided variant PAS. Using Pearson Chi-Square Test, a statistically significant ethnic variation in prevalence rates was identified at $p < 0.05$ (χ^2 (6, N = 1106) = 22.80, $p < 0.0001$). Asian subjects had the highest prevalence rate (6.6%), followed by African Americans (4.5%), Middle Easterners (3.4%), Mixed (2.4%), First Nations (2.0%), and Caucasians (1.2%). No PAS were found in South Asians or Latin Americans (see Table 4).

3.3. Family history

55 subjects (5.1%) out of the 1073 subjects able to complete the family history portion of the survey were found to have a positive family history, defined as the presence of a PAS in a first-degree relative. Of the 26 subjects positive for a PAS, 11 (42.3%) were found to have a positive family history. More specifically, 4 of the 7 (57.1%) subjects positive for bilateral PAS had a positive family history, and 7 of the 19 (36.8%) subjects positive for unilateral PAS had a positive family history. The odds ratio for a positive family history and the presence of a PAS was statistically significant at $p < 0.05$ (OR = 16.7, 95% CI: 7.3–38.5, $p < 0.0001$). The odds ratio for a positive family history and bilateral PAS was also statistically significant at $p < 0.05$ (OR = 26.5, 95% CI: 5.8-121.7, $p < 0.0001$) and greater than that seen in unilateral PAS (OR = 12.2, 95% CI: 4.6-32.5, $p < 0.0001$).