Published in final edited form as:

J Child Neurol. 2016 March ; 31(4): 468–473. doi:10.1177/0883073815599261.

Prevalence and Clinical Characteristics of Headaches in PHACE Syndrome

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Abstract

PHACE syndrome is a neurocutaneous disorder often involving the cerebral vasculature. PHACE patients appear to have early-onset and severe headaches more commonly than children without PHACE. The objective of this study was to characterize the clinical features and prevalence of headache by conducting a cross-sectional survey of families in two large PHACE registries. Sixty-six percent of eligible families completed the survey in which 62.7% of respondents reported headaches. Average age of headache onset was 48.8 months. Females were more likely to have headaches (68.6% versus 30.8%, $p=.014$). Families reported associated migrainous features including nausea (62.5%), vomiting (37.5%), photophobia (75%), and phonophobia (75%). Headaches occurred at least weekly in 29.4%, lasted ≥ 1 hour in 85.4%, and led to ≥ 1 hospital admission in 15.7%. Three respondents with headaches had at least one ischemic stroke. We demonstrated that headaches are common among PHACE patients, develop at an early age, and have migrainous features.

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Author Contributions

JY and GLH designed the study, collected the data, analyzed the findings, and prepared the final manuscript. DHS and BAD assisted in study design, collection and analysis of the data, and manuscript preparation. FB, LGE, and DM helped in data collection and manuscript preparation.

Ethical Approval

The institutional review board at the Children's Hospital of Wisconsin approved this study.

Declaration of Conflicting Interests

The authors declare no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Keywords

Headache; PHACE; Pediatric; Adolescent; Migraine; Pascual-Castroviejo Syndrome Type II; Hemangioma; Vasculopathy

Introduction

Infantile hemangiomas (IH) are the most common benign vascular tumors affecting 2.5%–21.0% of all newborns. Approximately 30% of newborns with large, segmental hemangiomas of the head and neck meet the diagnostic criteria for PHACE syndrome¹ (Online Mendelian Inheritance in Man database No. 606519). Coined by Dr. Ilona Frieden in 1996², the PHACE acronym summarizes the cardinal features of the syndrome: *p*osterior fossa brain malformation, *h*emangiomas, *a*rtorial anomalies, *c*oarctation of the aorta and cardiac defects, and *e*ye abnormalities.

Our PHACE syndrome registry and published case series have demonstrated that headache is a common symptom among PHACE patients^{1,3–6}. Some patients and families report debilitating headache episodes. While research data are lacking, clinical experience suggests that PHACE patients have headaches that are frequent and severe, begin at an early age, and cause considerable health-related disability. How PHACE syndrome influences headache risk is largely unknown. The high rates of cerebral vascular anomalies (seen in 91% of patients⁷) and structural brain lesions (seen in 42% of patients³) may predispose to higher headache rates. To date, no studies exist that have assessed headache characteristics from a large cohort of PHACE patients.

The PHACE Syndrome International Registry and Genetic Repository at the Medical College of Wisconsin constitutes the largest existing PHACE syndrome cohort representing a range of disease severities and diverse demographic features.

The aim of the current study was to characterize headache prevalence and the clinical headache features from a large PHACE syndrome cohort. We hypothesize that patients with PHACE syndrome will have high overall headache rates, severe headache episodes, and relatively young ages of onset.

Methods

We conducted a cross-sectional survey of patients with PHACE syndrome or their parents from December 2013 to June 2014. We identified patients through their enrollment in the PHACE Syndrome International Registry and Genetic Repository. Only patients recognized as having definite PHACE syndrome were included in the analysis.

An Internet-based survey was created with some questions adapted from the previously validated Diagnostic Interview of Headache Syndromes-Child version (DIHS-C)⁸ and some questions derived from the International Classification of Headache Disorders-3 (ICHD-3)⁹ diagnostic criteria. The headache disability questions were incorporated from the Pediatric Migraine Disability Assessment (PedMIDAS), which measures 3-month headache disability

in school-aged children.¹⁰ Each eligible family was sent an introductory e-mail that included a unique survey link. Up to three reminder emails were sent. Families received a gift card of small monetary value after completing the survey.

Ethics:

The Institutional Review Board at the Children's Hospital of Wisconsin and the Vascular Birthmark Institute of New York approved this study.

Data Analyses.

Data were included in the analysis if the parent or patient completed enough of the survey to document the presence or absence of headaches. Descriptive statistics were calculated. Categorical variables were compared using the Pearson chi-square test. Comparisons of continuous variables were made using the Student's t test or a corresponding non-parametric test. All statistics were performed using SPSS Version 21 (SPSS Inc., Chicago, IL, USA). Statistical significance was set at 5%.

Results

Among 200 families in the PHACE Syndrome International Registry, 144 had updated contact information and were eligible. Of those, 95 (66%) completed the survey. One patient denied having PHACE syndrome, and 11 parents did not know if their child had headaches because of young age or cognitive impairments. Responses from the remaining 83 surveys were analyzed.

The mean age of the 83 analyzed patients was 97.1 months (range 6–413 months). There was a female predominance (84.3%), which mirrors the high female-to-male ratio in PHACE syndrome. Fifty-two (62.7%) patients reported headaches. Headache patients were older than non-headache patients (mean 119.2 months versus 60.1 months, $p < .001$). Females were more likely to have headaches than males (68.6% versus 30.8%, $p = .014$). Family history of at least one first-degree relative with headache did not differ between headache and non-headache patients (59.6% versus 48.4%, $p = .32$).

Specific headache characteristics varied among patients (Table 1). Headache onset occurred at an early age (mean 48.8 months, range 12–130 months). Headaches occurred at least once weekly in 29.4% and lasted ≥ 1 hour in 85.4%. Several families reported associated migrainous symptoms including nausea (62.5%), vomiting (37.5%), photophobia (75%), and phonophobia (75%). Visual disturbances and osmophobia were acknowledged during headache episodes by 27.1% and 20.8% of families respectively. Fifteen patients (30.6%) reported having had auras or premonitory symptoms that preceded one or more headaches; 5.8% had an aura or premonitory with every headache episode. Since parents answered survey questions for the younger patients, we did not make further attempts to distinguish prodromal symptoms from migraine auras. Aura/premonitory symptoms included an "ill-feeling" in eight (15.4%) and visual changes in six (11.5%) patients prior to the onset of head pain.

Nine families reported a history of stroke or stroke-like episodes. Three patients had at least one arterial ischemic stroke, presumably related to PHACE vasculopathy; two reported a stroke *in-utero*; two had strokes related to surgical procedures; and two reported having stroke-like episodes and transient ischemic attacks. All patients were female, and all reported headaches. Overall, stroke patients had a lower mean headache intensity (3.57 versus 5.78, $p = 0.03$) and an older age at headache onset (62 months versus 46.6 months, $p = 0.035$) than PHACE patients without prior stroke.

Table 2 summarizes the clinical evaluations and treatments for headache patients. Over half (55.1%) of the patients had consulted with a physician about headaches and received diagnoses, which included migraine (55.6%), primary headache other than migraine (18.5%), hydrocephalus (3.7%), and moyamoya syndrome (3.7%). Five patients (18.5%) who consulted with a physician did not receive a headache diagnosis or the families did not know the diagnosis. Eight (15.7%) patients had been admitted to hospital for headache treatment or headache evaluation, and 52.2% had been seen by a school nurse for headache complaints. Non-prescription analgesics had been used by the majority of patients (89.8%), starting at a mean age of 4.8 years. Fewer patients (18.4%) used prescription analgesics, starting at a mean age of 7.8 years. Five patients (10.2%) used daily prescription medicines for headache prophylaxis.

PedMIDAS questions were used to assess headache disability at the time of survey, although the scale is valid for school age children only. After excluding patients younger than 5 and older than 18 (school-aged), 36 subjects remained. Thirteen (36.1%) reported no disability (PedMIDAS score 1–10); thirteen (36.1%) experienced some disability (PedMIDAS score 11–30); five (13.9%) had mild to moderate disability (PedMIDAS score 31–50) and 5 (13.9%) had severe disability (PedMIDAS score >50). Eighteen (50%) reported disability related to daily activities, 19 (52.7%) reported disability related to activities at home, and 13 (36.1%) reported disability at school.

Discussion

We conducted a survey of the clinical headache features from the largest PHACE cohort in the world. Our results demonstrate that headaches are common among patients with PHACE syndrome. The clinical characteristics vary. Many patients develop headaches at a relatively early age, several have associated migrainous symptoms, and some have a severe headache burden. Some PHACE patients may have concurrent primary and secondary headaches.

The prevalence of primary headaches in children varies by age and by the headache criteria used. Within the first three years of life an estimated 3–8% of healthy children will have experienced some form of headache.^{11,12} The rate increases progressively with age. By 7 to 15 years, 57–82% of children will have complained of at least one headache episode.^{13,14} Applying International Headache Society (IHS) diagnostic criteria, the prevalence of migraine in children 5–15 years of age has been estimated at 10.6%.¹⁵ An analysis of population-based studies that apply IHS criteria for migraine and tension-type headache estimates the mean prevalence of all primary headaches in children and adolescents to be 51%, while the prevalence of pediatric migraine is estimated at 7%.¹⁶ In the present study,

we demonstrate that 55.3% of patients from a large PHACE syndrome cohort suffer from headaches. While our exploratory research findings cannot be compared directly to historical headache rates in healthy children, headache appears to be a common symptom among patients with PHACE syndrome. The mean age of headache onset in our cohort was low compared with the general population, which has been reported at 9.7 years.¹⁷ Patients with headaches in the present study were older than patients without headache. The young age of some patients in our survey, may have led to an underestimation of the overall headache prevalence with this neurocutaneous syndrome. There is a female predominance in PHACE syndrome. Interestingly, the females in our survey were also more likely to develop headaches than the males, a phenomenon seen after puberty in primary headache epidemiology studies.

Headache constituted a substantial medical burden in our cohort. The World Health Organization has ranked headaches among the top most disabling medical conditions.¹⁸ Headaches in children can lead to reduced participation in school and social activities and to a negative impact on quality of life.¹⁹ We used the PedMIDAS to demonstrate the impact of headaches on school, family, and social domains in our cohort.¹⁰ Some of our surveyed patients were not school age, so the PedMIDAS did not adequately measure their headache-related disability. Additionally, many of the patients surveyed had been evaluated by a physician because of headaches. Children with a chronic illness such as PHACE syndrome may be more likely to consult a subspecialist for headaches, especially given the specific concerns about cerebral vascular changes or structural brain problems with this disorder.

A large proportion of the patients with headaches reported concomitant migraine symptoms including nausea, vomiting, photophobia, and phonophobia. Migraine aura can be a difficult symptom to define by survey, especially when parents answer the questions. A relatively high number of respondents acknowledged auras or premonitory symptoms (26.5%), described as an “ill feeling” or a visual disturbance that precedes headaches. In contrast, the estimated prevalence of migraine aura in adults is only 5%, with most patients experiencing visual phenomena and 54% describing sensory changes.^{20,21} Many of the clinical features in our PHACE cohort can be consistent with the IHS diagnostic criteria for migraine headaches (Table 3).⁹ However, we are cautious about labeling headaches as primary migraines when arterial anomalies and structural brain lesions are prevalent in this population.

Arterial anomalies of the head and neck are the most common (extra-cutaneous) feature of PHACE syndrome affecting up to 91% of all patients.²² Pleurisegmental arterial dysgenesis can lead to vascular looping, elongation, ectasia, kinking, and aneurysmal enlargement, particularly in the large vessels such as the internal carotid artery.⁷ Regional changes in cerebral blood flow due to arterial irregularities are thought to trigger cortical spreading depression in non-PHACE patients.²³ Cucchiara and colleagues have shown that migraineurs, especially those with aura, were significantly more likely to have an incomplete Circle of Willis than the general population.²⁴ This is relevant to the present study because patients with PHACE syndrome have higher numbers of arterial anomalies in general and a higher number variations of the Circle of Willis (absence, hypoplasia, and duplication of the anterior and posterior communicating arteries, middle cerebral artery, and anterior cerebral artery) compared to non-PHACE patients which may contribute to the headache

pathophysiology with this disorder.¹ Progressive cerebral vascular changes could also increase headache rates among PHACE patients. For example, moyamoya vasculopathy, characterized by cerebral vascular stenoses and the proliferation of small collateral vessels, occurs much more commonly in PHACE syndrome than in the general population.^{25,26} Moyamoya can present with migraine-like headaches related to cerebral hypoperfusion.

Cardiac abnormalities including aortic arch defects, structural heart defects, and aberrant origin of vessels are seen in 41% of PHACE patients.²⁷ In these patients with cardiac abnormalities, brachiocephalic abnormalities and coarctation of the aorta are the most common seen in 56% and 45%, respectively. The most common cardiac abnormalities in PHACE syndrome include ventral septal defects (VSD) and patent foramen ovale (PFO).²⁷ PFO is more common in adult and child migraineurs with aura than in the general population.²⁸ Closure of PFOs has led to improvements, and even resolution, of migraine attacks in some patients.^{29,30} Though the pathophysiology is not clear, the PFO may trigger migraines by allowing vasoactive substances to bypass the pulmonary circulation to reach the brain.³¹ The high prevalence of cardiac abnormalities, including PFO, in PHACE patients may also add to headache risk.

There are several limitations in the present study. Surveys do not capture objective data and are prone to recall biases. Headache prevalence in children increases with age, so inclusion of younger children in this survey may have led to an underestimation of the overall headache rates with PHACE syndrome. Parents served as survey respondents for many of the PHACE patients, which may have led to incorrect headache reporting. Nakamura and colleagues have shown that agreement between parent and child about the mere presence or absence of headaches can be very low.³² The stroke data is taken from the survey and unconfirmed with medical records and therefore subject to incorrect reporting. Finally, though our survey response rate was excellent, families whose children have more severe headaches may be more likely to respond to a survey about headaches, potentially skewing the data.

In conclusion, the present study shows that headaches are prevalent among PHACE patients. The headaches can be severe, often have associated migraine symptoms, and can cause substantial headache-related disability. Future study of headache pathogenesis and correlation with cerebral vascular and cardiovascular anomalies in PHACE syndrome is warranted.

Acknowledgement:

This work was performed at the Medical College of Wisconsin and the Children's Hospital of Wisconsin in Milwaukee, Wisconsin. The authors acknowledge Shawna Joachim (Medical College of Wisconsin) for her assistance with IRB approval, editing, and submission assistance. We want to thank all the patients and their respective families for responding to our survey and to helping further our understanding of PHACE syndrome. We would also like to thank the PHACE Syndrome Community for their continued support for our research and finally, we would like to thank the NIH for their generous grant support and the Greater Milwaukee Foundation for supporting this study.

Funding

Funding for this study was provided by the National Institute of Health (R01 AR064258-01A1).

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Table 1:**Headache characteristics**

PHACE Patients with Headache, n	52
Mean age at survey completion (months)	119.7
Mean age at headache onset (months)	48.8
Headache Burden, n = 51	
>20 headaches in lifetime	27 (52.9%)
At least 1 headache in past month	23 (44.2%)
At least 1 headache in past year	45 (86.5%)
Headache Location, n = 51	
Frontal (front of head)	23 (45.1%)
Multiple locations	17 (33.3%)
Temporal (side of head)	6 (11.8%)
Other/unknown	3 (5.9%)
Occipital (back of head)	2 (3.9%)
Headache Frequency, n = 51	
Daily	2 (3.9%)
Several days per week	6 (11.8%)
Once weekly	7 (13.7%)
1–3 times monthly	12 (23.5%)
Less than once monthly	24 (47.1%)
Headache Quality, n = 51	
Throbbing/pounding	17 (33.3%)
Aching/tightness	10 (19.6%)
Stabbing	4 (7.8%)
Other/unknown	20 (39.2%)
Duration, n = 48	
<1 hour	7 (14.6%)
1–4 hours	22 (45.8%)
4–24 hours	16 (33.3%)
>24 hours	3 (6.3%)
Headache Intensity (1–10 scale), n = 51	
Median	5
Associated Symptoms, n = 48	
Photophobia (aversion to bright lights)	36 (75%)
Phonophobia (aversion to loud noise)	36 (75%)
Nausea	30 (62.5%)
Vomiting	18 (37.5%)
Vision disturbance	13 (27.1%)
Osmophobia (aversion to strong smells)	10 (20.8%)

Aura or premonitory symptoms, n = 49	
Presence of any aura or headache prodrome	15 (30.6%)
Aura/prodrome with every headache	3 (5.8%)
Aura/prodrome with some headaches	12 (24.5%)
Visual aura	6 (12.2%)
Tingling or numbness aura	1 (1.9%)
Ill sensation or other sensation as aura	8 (16.3%)

Table 2:

PHACE patient headache evaluations and treatment

PHACE patients with headaches	52
Patient has gone to the school nurse for headaches (n = 46)	24 (52.2%)
Patient has gone to the hospital for headaches (n = 49)	8 (15.7%)
Patient has been evaluated by a physician for headaches (n = 49)	27 (55.1%)
Physician diagnoses based on respondent report (n = 27)	
Migraine headaches	15 (55.6%)
Non-migraine headaches	5 (18.5%)
Headache due to hydrocephalus	1 (3.7%)
Headaches due to moyamoya vasculopathy	1 (3.7%)
Unknown	5 (18.5%)
Has used non-prescription analgesics (n = 49)	44 (89.8%)
Mean age in years, non-prescription analgesics started	4.8
Non-prescription analgesics are effective	32 (65.3%)
Has used prescription headache medicines (n = 49)	9 (18.4%)
Mean age in years, prescription headache medicine started	7.8
Daily (prophylactic) prescription headache medicine	5 (10.2%)

Table 3:

Diagnostic criteria for migraines in children (less than 18 years of age)

ICHD-3 Diagnostic Criteria for Migraine Headaches in Children < 18 Years of Age	
A. At least 5 attacks fulfilling criteria B-D	
B. Headaches lasting 2–72 hours	
C. Headaches with at least two of the following four characteristics	
1	Unilateral or bilateral (more common in children) location
2	Pulsating quality
3	Moderate or severe pain intensity
4	Aggravation by or causing avoidance of routine physical activity
D. During headaches at least one of the following	
1	Nausea and/or vomiting
2	Photophobia and phonophobia
E. Not better accounted for by another ICHD-3 diagnosis	