180 Burden of Hereditary Angioedema: Findings From a US Patient Survey

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RATIONALE: Hereditary angioedema (HAE) is characterized by unpredictable and potentially severe or life threatening attacks that negatively impact daily life. With several novel acute and preventive HAE treatments available, the impact of HAE on health-related quality of life (HRQoL) is of interest.

METHODS: A web-based survey of randomly selected members of the US Hereditary Angioedema Association was conducted (March through April, 2017). Respondents were ≥18 years old, had type I/II HAE, ≥1 attack during the last year, and received an HAE treatment within the last 2 years. HRQoL was measured using HAE-QoL and SF-12 questionnaires. Additionally, the Hospital Anxiety and Depression Scale (HADS) and the Work Productivity and Activity Impairment (WPAI) questionnaire were used. Descriptive analyses were conducted.

RESULTS: Of 707 patients screened, 445 (62.9%) completed the survey. Most were 18-64 years (92.8%), female (78.2%), and 68.5% received prophylactic treatment. Mean (SD) number of attacks was 11.1 (13.9) during the past 6-months and 78.7% reported an attack during the past month. HAE-QoL scores were highest (better QoL) among those reporting 0-1 attacks during the past 6-months, and decreased at higher attack frequency. Highest impairment (lowest scores relative to maximum) was reported in the ‘perceived control over illness’ domain. Mean (SD) SF12 mental and physical composite scores were 44.9(10.9) and 48.6(9.9), respectively. Respondents reported mild-to-severe anxiety (49.9%) and depression (24.0%). Mean (SD) percentage impairment measured by WPAI was 25.4(28.1) for work-productivity loss and 31.8(29.7) for activity impairment.

CONCLUSIONS: In this study, patients with HAE reported anxiety and depression and impairments in HRQoL and work productivity.

181 Is There a Difference in Omalizumab Treatment Response in Patients With Chronic Autoimmune Urticaria vs Chronic Spontaneous Urticaria?

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RATIONALE: To investigate the correlation between utility of chronic urticaria index (CUI), concurrent use of systemic steroids, antihistamines, and persistence of symptoms in patients with refractory chronic idiopathic urticaria (CUI), treated with omalizumab.

METHODS: Retrospective chart review of 39 patients with either Chronic Autoimmune Urticaria (CAU) or Chronic Spontaneous Urticaria (CSU) treated with omalizumab in a university based practice. Histories were evaluated for resolution of symptoms, prednisone use, CUI, and dose related side effects to either 150 mg or 300 mg of omalizumab.

RESULTS: Of the 39 patients, thirty (76%) had CUI drawn, eleven (36%) of which were positive. Thirty- two patients (82%)were on 300 mg while only seven (17%) were on 150 mg of omalizumab. Patients who experienced hives prior to their injection on week four were switched to a three week regimen with improvement in symptoms. Two patients on 300 mg experienced headaches and dizziness. One had their dose decreased to 150 mg with resolution of symptoms, the other discontinued the treatment. Twenty-four of the thirty-nine patients (61%) on prednisone prior to starting omalizumab no longer required prednisone.

CONCLUSIONS: When comparing CAU and CSU patients on Omalizumab, we found no difference in the symptom resolution or corticosteroid use. However, omalizumab was an effective steroid sparing agent overall. Rather than discontinuing omalizumab all together due to certain side effects, dose reduction may help to resolve adverse effects while still maintaining control. In addition, an increase in dose frequency may help to maintain symptom control.

182 The Predictability of Attacks in Patients with Hereditary Angioedema

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RATIONALE: Patients with hereditary angioedema (HAE) have debilitating attacks of swelling. We conducted a systematic literature review to evaluate patients’ differing ability to predict the onset of these attacks and the consequences for quality of life.

METHODS: The literature search of PubMed consisted of the following terms and their variations: trigger, prodromal symptom, predict, variable, breakthrough, fear, functioning, activities, daily life, spontaneous, travel, anxiety, impairment, certainty, productivity, absenteeism, work, future attack, worry, and episodic. Article abstracts were reviewed for relevance to the predictability of attacks in patients with HAE.

RESULTS: The search returned 312 articles, of which 79 were identified to be potentially relevant to the predictability of HAE. In two studies that asked patients about the predictive utility of prodromal symptoms, most patients were not able to predict attacks reliably. However, 85% and 91% of patients, respectively, stated that attacks were preceded by prodromal symptoms at least half of the time. No studies prospectively evaluated the positive predictive value of prodromal symptoms. Attacks are not predictable over the long term, which causes anxiety about future attacks and impairs patients’ ability to maintain employment, be productive, and conduct daily activities.

CONCLUSIONS: Patients can predict some attacks over the short term using prodromal symptoms, but the prevalence and characteristics of prodromes are highly variable. HAE symptoms are not predictable over the long term, which negatively affects patients’ quality of life. The unpredictability of HAE symptoms should be considered when establishing HAE treatment plans.

183 Effect of Antileukotriene Agents in Chronic Urticaria-Isolated Angioedema Phenotype

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RATIONALE: Ten percent of patients with chronic spontaneous urticaria (CSU) have isolated angioedema. Although, antileukotriene agents are beneficial in NSAID-intolerant CSU patients, the evidence for benefit in other CSU patients is mixed. We assessed the efficacy of antileukotrienes in CSU-isolated angioedema phenotype.

METHODS: Retrospective review of CSU-isolated angioedema patients evaluated at Johns Hopkins Allergy Clinic between October 2008 and April 2017. Each patient’s symptoms before and after LTRA treatment was graded on a 7-point scale based on frequency and severity of episodes. RESULTS: 46 patients were identified and 17 were lost to follow-up after the initial visit. Of the 29 patients included in the study, mean age was 57 years, and 16 (55%) were female. 24 (83%) had received antileukotriene agent in conjunction with antihistamines. 17 (71%) of these 24 patients had sought urgent treatment for an episode of swelling while 2 (7%) had been intubated due to swelling prior to the initial visit. 22 (92%) had significant improvement in their symptoms following addition of anti-leukotriene agent (p<0.00001) with 15 (62.5%) having had complete resolution of symptoms over the follow-up period ranging 4 months to 7 years. Of the 22 who had response to antileukotriene agent, only 5 (22%) had reported NSAID-intolerance. 1 of the patients with no response to antileukotriene had response to doxepin while the other was placed on anti-immunoglobulin E therapy.

CONCLUSIONS: In patients with CSU-isolated angioedema, antileukotriene therapy appear to control symptoms when added to anti-histamines. However, larger prospective studies are needed to validate these results and identify predictive markers.