

HAE in Australia in 2015: results of HAE Australasia survey.

Although the situation for those living with HAE has improved, the recent HAE Australasia Patient Survey reveals the continuing burden of living with this condition. Twenty two participants at the annual conference in May 2015 gave consent and completed a questionnaire. As this year's meeting was held in Sydney, it was to be expected that the majority of those answering the questions lived in NSW or Victoria. Not surprisingly, the majority had other family members affected by this genetic disorder; only 4/22 had no family history of HAE.

Nearly half those responding to the survey stated that their diagnosis was made because of an affected family member; one "lucky" person was diagnosed correctly after the very first angioedema attack; eleven people waited many years for a correct diagnosis.

HAE attacks are very debilitating and interfere with daily life. Half the survey group stated they experienced one attack per month on average, while six people experienced more than one attack a fortnight and one person averaged two attacks per week. In the last 12 months, most people had missed days from work as a direct consequence of HAE; seven respondents had lost 5 days and only one had not missed work because of HAE .

The advent of self management and the availability of "on demand" treatment has definitely impacted positively on the lives and well being of those living with HAE and this is reflected in the marked reduction in the number of visits needed to the emergency department in the last 12 months. Only five people had made more than

one visit to an ED; eight had not had any visits. This is in marked contrast to the results of the first HAE survey in Australia where 50% of people had experienced 10 or more visits to an ED.

Those living with HAE continue to nominate “stress” as the single most common cause precipitating an attack (15/22); trauma was nominated by 14; infection by eight and menstrual period a trigger in five people. One third of those answering the questionnaire volunteered depression and anxiety as a co morbidity. According to Sane Australia, during a one-year period, anxiety disorders will affect 14% of the population and depression will affect 6%, thus those living with the burden of HAE have a much greater risk of these debilitating disorders and this gives HAE Australasia an opportunity to explore interventions to alleviate this burden among its members.

The survey explored members’ use of HAE medication. Both tranexamic acid (50%) and danazol (75%) had been used by many respondents. Not surprisingly in this adult population, most had found danazol more effective than tranexamic acid.

Firazyr (icatibant) had been used by 15 of 19 who replied to this question; one person had not found it effective and one was unsure regarding its effectiveness but all others had found it effective. In the last 6 months, the number of injections used ranged from once (3 respondents) to more than five times (8 respondents) and all stated that having this medication had prevented hospitalisation.

C1inhibitor concentrate (Berinert or Cinryze) had been accessed by 9/19 respondents, four of these occasions had been for pre-operative prophylaxis.

This survey, as did the previous one of Australians living with HAE, has shed light on the continuing burden of having this disorder and while the situation has definitely improved, there is still more to be done.

Finally, we thank all those who took the time to participate in this survey. It is only by knowing about the burden faced by those with HAE and their continuing difficulties that we can hope to advocate for improvements in management.

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