IMPACT OF MENSTRUAL CYCLICITY SUPPRESSION ON SEVERITY AND FREQUENCY OF SYMPTOMS OF HEREDITARY ANGIOEDEMA IN A PAEDIACTRIC GYNAECEOLOGICAL POPULATION

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INTRODUCTION

Hereditary angioedema (HAE) is a rare autosomal dominant condition that affects 1:50,000 individuals and is primarily due to C1-inhibitor dysfunction resulting in recurrent attacks causing inflammation and swelling of subcutaneous or submucosal tissue. HAE can be debilitating to patients due to cyclicity of symptoms with triggers such as menstrual cycles.

OBJECTIVE

This original research explores menstrual cyclicity suppression with hormonal treatment using progesterone alone on the frequency and severity of HAE attacks in a paediatric gynaecological population

METHODOLOGY

7 patients with HAE were known to paediatric gynaecology at a tertiary hospital and included in this original research. Data were collected through physical and electronic patient records.

Ethics approval was obtained from the ethics committee at the Royal Bribane and Women's Hospital.

RESULTS

The most common trigger for attacks in 71.43% of patients were menstrual cycles and oestrogen. Although, there were numerous reported triggers as demonstrated in Table 1.

The severity of symptoms varied between patients as demonstrated in Table 2. Severe cases included debilitating symptoms of vulval oedema resulting in menses obstruction secondary to swelling. 28.57% of patients required intensive care admission secondary to attacks. One patient had prophylactic monitoring of each menses before treatment due to recurrent intubation secondary to laryngeal oedema.

The duration of symptoms prior to treatment exhibited variability. 80% of patients who experienced menses as a recognised trigger, the attacks persisted throughout the entire duration of their menstrual cycle. One patient did not have any identifiable triggers for HAE and encountered a daily recurrence of symptoms that never fully subsided

HAE attacks were cyclical for patients who had menses as a known trigger resulting in recurrent adverse effects to their quality of life. The average age of patients was 14.7 years old, and recurred attacks resulted in decreased participation in school and ability to complete daily activities and hobbies. 28.57% of patients required support from mental health care teams.

71.42% of HAE patients were over-investigated with additional imaging or laparoscopic surgery, which did not yield any significant findings to elucidate their symptoms.

Management of HAE triggers was completed by menstrual suppression with progesterone-only pills, depot provera intramuscular injection (IMI), and Mirena intrauterine device (IUD). 85.71% of patients achieved a reduction in the cyclicity and severity of symptoms with treatment resulting in improvement of quality of life. One patient did not have improvement and experienced stomach upset, laryngitis, and post-nasal drip secondary to the commencement of treatment with Depot Provera but failed to follow up in the clinic for further treatment adjustment.

71.43% of patients were initially trialled on progesterone-only pills however 80% of those patients were then transitioned to either the depot provera IMI or Mirena IUD due to adverse effects experienced with attack recurrence with breakthrough bleeding. The Depot Provera IMI and Mirena IUD were the most effective options. 100% of patients tiralled on the Mirena IUD and 75% of patients tiralled on the Depot Provera IMI achieved treatment success with a reduction in cyclicity and duration of attacks alongside a reduction in symptom severity and an overall improvement in their quality of life.

Immunology specialists were engaged in the care of 71.43% of patients and provided respective treatment. For all patients, immunology teams supported treatment success achieved with menstrual impression and recommended continuing management with menstrual suppression.

Table 1: Triggers for HAE patients

	Menstural Cycle/ Oestrogen	Procedures	Stress	Other
Patient 1	Yes	Yes	Yes	Weather Changes
Patient 2	No	Yes	Yes	Illness
Patient 3	Yes	No	No	
Patient 4	Yes	No	No	Aeorosoles
Patient 5	Yes	No	No	Warm water
Patient 6	Yes	No	No	
Patient 7	No	No	No	
Total n (%)	5 (71.43%)	2 (28.57%)	2 (28.57%)	

Table 2: Symptoms experienced during HAE attacks

Symptom	Description	Number of patients (%)
Blood pressure-associated symptoms	Hypotension and Collapse	3 (42.86%)
Skin and mucosal involvement	Angioedema of the face and limbs, Urticarial rashes , Vulval oedema, Laryngeal oedema, Puritis	7 (100%)
Gastrointestinal symptoms	Abdominal Pain , Bloating, Nausea and vomiting,, Constipation, Diarrohea	7 (100%)
Other	Breast tenderness, Bronchospasm, Laryngospasm , Seizure like activity, Hot flushes, Shortness of breath	5 (71.43%)

DISCUSSION

Most patients were initially trialled on a short-acting reversible management option with the progesterone-only pill likely as the potential adverse effects to HAE attacks were unknown.

Management of HAE involves multidisciplinary care and strong co-accordance between teams including immunology and mental health should be supported in earlier for care planning.

There was variable documentation in clinic notes due to clinician variability resulting in discrepancy in documentation of HAE attack symptomatology that could be standardised for further research

CONCLUSION

HAE management of patients with menses as a known trigger was most effectively achieved with the Depot Provera IMI and Mirena IUD resulting in a reduction of the cyclicity and severity of symptoms. However, the care of patients with HAE requires multidisciplinary input to optimise the reduction of disease burden on patients. There is limited research in the area of HAE in an Australian Paediatric Gynaecological population and this original research should be extended with a larger cohort in the future.

REFERENCES

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