COMPLEMENT

C1 Esterase Inhibitor (C1-INH) deficiency is associated with hereditary angioedema.

Hereditary Angioedema (HA)

- ➔ usually starts in **childhood**
- ➔ **recurrent episodes** of angioedema and/or abdominal pain
- ➔ may involve the larynx therefore can be fatal
- ➔ diagnosis is suggested by recurrent episodes of non-urticarial swelling unresponsive to antihistamines, positive family history, episode of laryngeal oedema, recurrent episodes of unexplained abdominal pain and vomiting
- ➔ steroids and anti-histamines are not effective in attacks
- ➔ also consider if angioedema episodes are; drug induced (ACEI), allergic, acquired, idiopathic
- ➔ investigation involves C4 levels, C1-INH levels and C1 function if available
- ➔ results dictate subtype- type I, II, or III

If hereditary angioedema is diagnosed patients may need blood products for future episodes. Plasma derived C1-INH shortens attacks and acts in 30-60 minutes. Therefore other investigations are likely to be required: FBC, U+Es, LFTs, lipids, blood borne virus testing, urinalysis and liver/spleen USS.

Complement C3 and C4 can be used to monitor disease activity in SLE. Levels fall with disease activity due to complement consumption.

Issues regarding testing:

- Hereditary angioedema is unlikely to be diagnosed in primary care. There is no adult allergy or anaphylaxis service in Edinburgh. Consider whether these patients would be referred to dermatology. However C1 testing may be useful if there are recurrent or life threatening episodes as it could guide management in the future.

- This test should not normally be initiated by primary care. Its role in diagnosis and management is uncertain and as such it may be more appropriately used in secondary care.

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