periodic paralysis [created by Paul Young 29/11/07]

general

-the term periodic paralysis describes a group of rare primary inherited disorders that are mostly autosomal dominant and cause periodic weakness

pathogenesis

- in general the underlying abnormality is a defect in a skeletal muscle ion channel

clinical features

- symptoms begin early in life (before age 25) and follow rest or sleep rather than exertion
- alertness is completely preserved during attacks & power is normal between attacks

hypokalaemic form
- predominantly inherited but can also arise sporadically
- involvement of bulbar or respiratory muscles is rare but can occur
- hypokalaemia during attacks is generally mild but symptoms respond rapidly to potassium supplementation
- acetazolamide provides effective prophylaxis

hyperkalaemic form
- milder and almost always inherited
- virtually never requires intensive care
- responds to carbohydrate administration
- thiazides or acetazolamide provide effective prophylaxis

treatment

- usually successful in both preventing attacks and preventing chronic weakness which may develop after many years of attacks in untreated patients