Background
As a tertiary centre for paediatric neurology, Southampton accepts referrals from across the region. A proportion of these referrals are patients that have suffered severe brain injury. Consistent themes emerged in two of the cases admitted over the last year. Both patients developed symptoms of hypertension, hyperthermia, tachycardia, tachypnoea, diaphoresis, extensor posturing and agitation; a syndrome which has been described in the literature, as Paroxysmal Autonomic Instability with Dystonia (PAID).

Objective
To illustrate how high dose clonidine has been used, with good effect, for the treatment of PAID.

Methods
Two, 14 year old male, patients who presented with severe brain injury and were diagnosed with PAID are described. The causes of injury were, near drowning and a road traffic collision (pedestrian versus car).

Results
Patient 1
Despite dose titration with an ace inhibitor and a calcium channel blocker, the only medicine that controlled the patient’s symptoms proved to be high doses clonidine of up to 800 micrograms every two hours (recorded blood pressure was 142/94 before administration of this dose and 126/75 post dose). Observations, including four hourly blood pressures, were recorded during treatment and more frequently after a change in the dose. When weaning the clonidine, by administering the same dose at a decreased frequency rebound hypertension occurred. Thus, clonidine was weaned very slowly by reducing the dose and maintaining the frequency. During an acute episode of PAID, the patient had a heart rate of 100 bpm, a respiratory rate of 24 breaths/min and a blood pressure 170/111.

Patient 2
High dose clonidine treatment was started early with a similar outcome following the experience from the first patient.

Conclusion
The diagnostic criteria for PAID syndrome has been proposed by Blackman et al as severe brain injury accompanied by a temperature of at least 38.5°C, pulse of ≥130 bpm, a respiratory rate of ≥140 breaths/min, agitation, diaphoresis and dystonia. The duration is at least one episode per day for at least three days. It is important when considering the diagnosis of PAID that other possible conditions, warranting different treatments, are excluded.

It is thought that the features of PAID can be explained by dysfunction of the autonomic centres in the brain, mainly the thalamus and the hypothalamus. Another theory believed to be responsible for the hypertension, is the cortically provoked release of adrenomedullary catecholamines. Other pharmacological agents previously used in an attempt to treat PAID, are morphine and non-selective β-blockers. Both have proved less favourable treatment options since opiate withdrawal may provoke signs of PAID and β-blockers, although useful for treating hypertension, do not affect cholinergic signs (e.g. diaphoresis). Clonidine, an α2-adrenergic agonist, has three actions; it reduces blood pressure, stabilizes behaviour and sedates.

Increasing awareness of this syndrome and its treatment with clonidine, will allow early intervention, decrease stress and anxiety for the patient/carer and above all prevent a hypertensive or hyperthermic encephalopathy.

Reference