

Post-Operative Opioid use in a Child with Cockayne Syndrome

Dr I.Songaile (ST7 Anaesthetics) , Dr C.Watkinson (Consultant Paediatric Anaesthetist)

Description:

Cockayne syndrome is a rare autonomic recessive inherited disorder. The diagnosis depends on the presence of three signs: microcephaly, impaired postnatal growth and premature pathological aging. Other recognized features include photosensitivity, hearing loss, cataracts, retinal dystrophy, abnormal liver and renal function and developmental delay.

The Cockayne Syndrome Natural History Study (2016) made a number of recommendations regarding medication use. This included avoidance of metronidazole, which can cause fulminant liver failure, but also to exercise 'extra vigilance' when dosing opioids and sedatives. Their recommended starting dose of opioids is 1/3 of the standard dose, as children in the cohort often had an exaggerated response to these drugs.

Case:

A 10-year-old male with Cockayne syndrome underwent multiple bilateral lower limb soft tissue release surgery. Initially, epidural postoperative analgesia was planned to negate use of opioids, however the surgeon wanted the patient to undergo early active physiotherapy, so a modified analgesia technique was implemented.

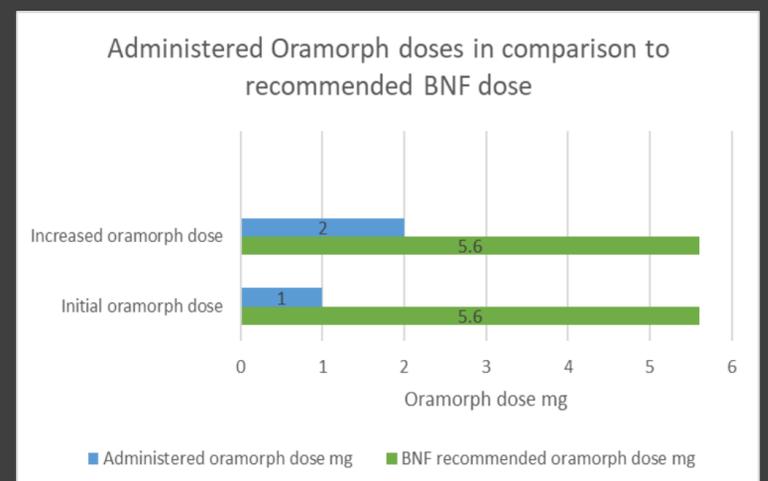
Following induction of general anaesthesia, the patient received caudal anaesthesia with 0.25% levobupivacaine and clonidine mix for perioperative and early postoperative pain relief. He was prescribed regular paracetamol and ibuprofen; 2 mg of diazepam twice daily as required for muscle spasm; and a reduced dose of oral morphine for breakthrough analgesia once the caudal analgesia wore off.

He weighed 28kg at that time of surgery. The BNF dose of oramorph is 200mcg/kg 4 hourly, 'adjusted to response.' We prescribed a third of a 100mcg/kg 2 hourly, as required. It may be questioned as to why we gave such a low dose initially – we felt it was better to have a low dose more frequently given the unpredictable way he might respond. But then it ended up being doubled (and actually given at 1/3 of the recommended dose), more frequently than 4-hourly.

No side effects were observed during and after the time period when oramorph was taken.

Discussion:

Adequate pain relief was achieved with the combination of simple analgesia, small dose of benzodiazepines and 1/3rd of recommended oramorph dose every 2 hours as required with no respiratory depression or sedative effect. When response to opioids is anticipated to be unpredictable, it is prudent to utilise opioid sparing techniques such as regional analgesia, regular simple analgesia, and analgesic adjuncts such as clonidine. This aims to ensure both patient safety and satisfaction



Analgesia during peri-operative and post-operative period



Reference:

Brian T. Wilson, Zornitza Stark, Ruth E. Sutton, Sumita Danda et al. The Cockayne Syndrome Natural History (CoSyNH) study: clinical findings in 102 individuals and recommendations for care. *Genetics in Medicine* 2016 18: 483–493