**Prescribing for children with adrenal disease**

Like Katharine Forward (Forward, 2015) I am a recently qualified Non-medical prescriber (NMP): I too am studying the Children’s Advanced Nurse Practitioner programme, but I am currently a Clinical Nurse Specialist in Endocrinology at a large children’s hospital in central London.

I have run nurse led clinics (NLC) previous Trusts I have worked in, but my Trust stipulate that to run a NLC, nurses must hold the NMP qualification, and also to have completed the Advanced Assessment in the Presenting Child module, of which I am near completion. Once the courses are completed (and passed!) a new nurse led clinic for managing children with adrenal disease will be born.

There are many types of adrenal disorders that we see here in our Trust: disorders of Glucocorticoid excess, such as Cushing’s syndrome, and Glucocorticoid deficiency. Cortisol deficiency is more commonly seen, and this could be secondary, caused by hypothalamic-pituitary disease, or primary, which can be congenital or acquired. The most common condition seen in the adrenal clinic is Congenital Adrenal Hyperplasia (CAH).

In CAH, 21-Hydroxylase deficiency is the most common type, with an incidence of around 1 in 15,000 births (Speiser et al., 2010). It an autosomal recessive condition, where deletions or mutations of the cytochrome P450 21 hydroxylase gene mean that there is glucocorticoid and mineralocorticoid deficiency (Hindmarsh, 2009). This is due to an enzymatic block (Trapp et al., 2011) in the pathway of steroid biosynthesis (White, 2001), and also results in over production of adrenal androgens due to the continued drive for ACTH production (Hughes, 2005), hence masculinised female genitalia. In male infants, diagnosis is not usually made until after day 7 of life, when a salt losing crisis with low plasma levels of sodium and high levels of potassium are indicative of diagnosis (Hindmarsh, 2009). Hence, babies with 21 hydroxylase deficiency CAH need to be started with glucocorticoids, fludrocortisone and also additional salt supplements (Raine et al., 2011). Girls with this type of CAH are usually identified at birth, and referrals to GOSH are made under the Disorders of Sex Development (DSD) team, for a multi-discplinary team review (Brain et al., 2010).

Whilst studying for the NMP qualification, I researched the prescribing for children with CAH in great detail. It is imperative to administer glucocorticoids to suppress the excessive secretion of corticotropin releasing hormone (CRH) from the hypothalamus, and ACTH by the anterior pituitary gland (Hindmarsh, 2009). This reduces the circulating concentration of androgens, which is especially important in females. Hydrocortisone is the glucocorticoid of choice because the risk of growth suppression in growing children is less severe (Trapp et al., 2011). Tablets are recommended at a dose of approximately 10-20mg/m²/day (Trapp et al., 2011, Raine et al., 2011, Speiser et al., 2010, Hindmarsh, 2009). In our centre, in infants, we prescribe 10-15mg/m²/day (Hindmarsh, 2009). Doses are spread 3 – 4 times throughout the day. Dosing as the child grows is titrated against body surface area calculations, 24 hour blood profile levels of cortisol and 17 hydroxyprogesterone, and side effects of under dosing (androgenisation) and overdosing (Cushing’s syndrome) are observed for.

I questioned the prescribing of Hydrocortisone tablets, as clearly administering a suspension of Hydrocortisone would be easier. Well, yes, but there is a good reason why not. While it may seem contradictory for an infant to be administered tablets, it has been proven that hydrocortisone suspension and hydrocortisone tablets are not bioequivalent in the treatment of children with CAH (Merke et al., 2001). Merke et al (2001) noted an increased need for higher hydrocortisone doses in their group of CAH children, resulting in an inadequate control of androgens, and signs and symptoms of Cushing’s syndrome. The research highlighted how the manufacturers of the suspension had changed the suspending agent in 1998. Following this research, the manufacturer withdrew their formulation from the market in 2000, but local hospital pharmacies still continue to formulate their own products. Such research was was also taken up by The Endocrine Society Clinical Practice Guideline (Speiser et al., 2010) and the guidance is utilised and cited internationally for best practice. However, watch this space for future changes, as difficulties in recent changed formulations of Hydrocortisone tablets are prompting prescribers to look again at suspensions..

With regards to my nurse led clinic: I hope to offer appointments to all newly diagnosed babies with CAH one month after diagnosis and discharge from our hospital. In between times, local community nursing teams take weekly samples of blood for U&Es, can liaise directly with me with the results, and doses hydrocortisone and salt supplements can be titrated by me if needed, over the phone to the parents. At the one month clinic appointment, further changes could be made, as well as re-iterating the importance of sick day and emergency management, but generally offering more support and guidance to the family as has previously been offered in the past. We offer a wide range of leaflets, support groups, and another CNS in our endocrine team has even have developed an app (MyCortisol) for families with emergency injection details. Subsequent visits will focus on issues regarding compliance, management of medication and dose changes, re-education of the condition and emergency management, arranging annual reviews, and also transition.

Qualifying as a NMP has really opened my eyes to the reality of prescribing in children, and has made me realise the importance of pharmacology and research with reference to different methods of drug formulations. It’s a tough course, but well worth it in the end, and I am very excited to be starting my adrenal nurse led clinic.

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