



USING THE PUMP METHOD IN CONGENITAL ADRENAL HYPERPLASIA

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*Health professional, parents, patients and families
working together to provide the best support for
those who have CAH*

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"The ideas expressed are independent of the editors' affiliations. Data provided is from current literature and should always be discussed with your endocrinologist"

Introduction

Mimicking the circadian rhythm of cortisol secretion is the ideal way of treating people with Congenital Adrenal Hyperplasia (CAH). We try to do this in a number of ways by giving hydrocortisone orally 3 or 4 times a day but although we try our best to get this as correct as possible it is not exactly the same as the normal circadian rhythm. However it is possible to mimic the circadian rhythm by using pump therapy. To see how hydrocortisone works when using it to replace cortisol, have a look at our hydrocortisone leaflet, where you will find examples of how the hydrocortisone (cortisol) works and its effects on the 17 OHP level, when taken twice a day, three times a day and four times a day.

<http://www.cahisus.co.uk/leaflets.htm> - HYDROCORTISONE

People with CAH can be both under and over treated even if their 17 OHP levels seem to be within normal range which is why having a full 24 hour profile to look at the cortisol is so valuable. Often patients complain of headaches, weight issues, tiredness and poor growth. These side effects can be a result of having periods of the day where they have too much cortisol and periods where no cortisol can be measured in the blood. To learn more about this please read our leaflet "The Value of 24 Hour Profiles".

<http://www.cahisus.co.uk/leaflets.htm> THE VALUE OF 24 HOUR PROFILES



Medtronic Pump



Animas pump



Coming soon the rechargeable, wireless and waterproof CellNovo Pump

How the Pump delivers

The pump method not only delivers the cortisol in a way which mimics the circadian rhythm, but also the way cortisol is naturally processed in the body, i.e. in the same way as a person who does not have CAH. In someone without CAH, cortisol is released directly from the adrenal glands into the blood stream and is then processed in the liver. When replacing cortisol in tablet form, the cortisol is first ingested into the stomach, processed through the liver and only then released into the blood stream. The pump method bypasses the stomach and you get 100% absorption straight into the blood stream.

The pump also allows us to adjust the dose accurately by very small amounts, at any time in the 24 hour period; ultimately we are able to set the rates to suit the individual's metabolism.

Examples

The following data are examples of two patients before using the pump method and then using the pump with the Peter Hindmarsh Formula.

Double and Triple Rates for Illness

Double and triple doses can be achieved very easily by switching over to the desired rate (these rates are worked out and programmed into the pump). Although the pump method also ensures the patient is getting cortisol during vomiting illnesses, it is still important to have the emergency injection on hand at all times and remember to increase fluid intake.

It is important when ill to give a bolus when switching over to double/triple rates; this gives the cortisol level an immediate boost.

Temporary Basal Function can be used to set a specific rate for a certain period of time. For example, the temporary basal rate can be set to deliver 150% which is the normal dose (100%) and an increase of half the normal dose (50%). The pump will deliver one and a half times the daily rate for as many hours as you set it to do this. This function is handy to use in exam situations or a situation where the patients feels they may need a little more cortisol, it is also very easily cancelled and the pump automatically reverts to deliver normal rates.

The pump gives the patient more independence as there is no forgetting or missing doses, no doses taken late, no having to wake up to take a dose and also no having to take a dose at inconvenient times.

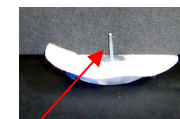
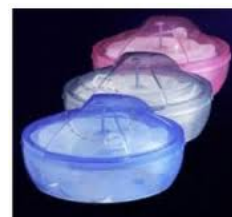
The pump is driven by batteries; the type depends on the manufacturer of the pump. Most Animas pumps use one AA lithium battery and most Medtronic pumps use two AAA lithium batteries. When using the pump for CAH, the batteries last several months and as soon as the pump senses a low battery an alarm alerts you to this.

When the reservoir is running low, the pump will alarm. You can see exactly how much the pump has delivered by looking at the history. Many pumps come with software, which allows you to download the delivery information.

If there is a problem with delivery, which is rare, the pump will alarm to alert you.

The Animas pumps are waterproof and can be used whilst doing water based activities such as swimming. All other pumps are very robust and are used by diabetics whilst doing all kinds of sport.

We recommend using Solu-Cortef in the potency 100 mgs to 1 ml of sterile water. For more information on how to do this, have a look at the pump video on our website



Automatic Inserters which are painless when used and leave the fine cannula in the body Watch our Pump Video on line <http://www.cahisus.co.uk>

The results in Figure: 5 show us: -

1. Despite the patient clearing cortisol rapidly the cortisol follows the circadian rhythm.
2. There are no periods where the patient is cortisol deficient.
3. The 17 OHP is suppressed to optimal levels.

An important factor to note is that is the overall daily dose of hydrocortisone delivered by the pump in the profile Figure: 5, is much lower than the overall daily oral dose taken in the profiles Figure: 3 and Figure: 4.

The patient's well-being and general health improved dramatically. The patient gained more height than expected and their weight issues resolved. The patients' BMI remains normal and their headaches, severe gastritis and lethargy disappeared. The patient also noticed a marked increased stamina when partaking in sport. Further studies showed that this patient had a problem with the cortisol-cortisone shuttle and by initially giving higher doses of hydrocortisone, it exacerbated the problem, the pump bypassed this issue.

Conclusion

The primary aim in treating Congenital Adrenal Hyperplasia is to replace cortisol and the 17OHP levels guide us to how much cortisol is needed. Hydrocortisone is a synthetic form of cortisol and it is the mildest steroid that can be used to replace cortisol. It is important to try to get the dose as accurate as possible in order to avoid the side-effects caused by replacing with either too much or too little cortisol as this can cause long term damage to the body.

In CAH it is also important to catch the early morning rise of ACTH to stop the over production of androgens, using the pump method achieves this. The body naturally produces cortisol in the early hours of the morning and the reason for this is thought to be because it is a period where no food or drink is consumed, so it is the body's natural way of keeping blood glucose levels normal.

As we can see from the profiles in Figure: 2 and Figure: 5, whilst using the pump with the Peter Hindmarsh Formula, the pump method can achieve perfect cortisol replacement on the minimum dose of hydrocortisone. However it is important that the rates are worked out for each individual patient as the pump can only deliver what it is programmed to do and does not have generic rates.

General Information

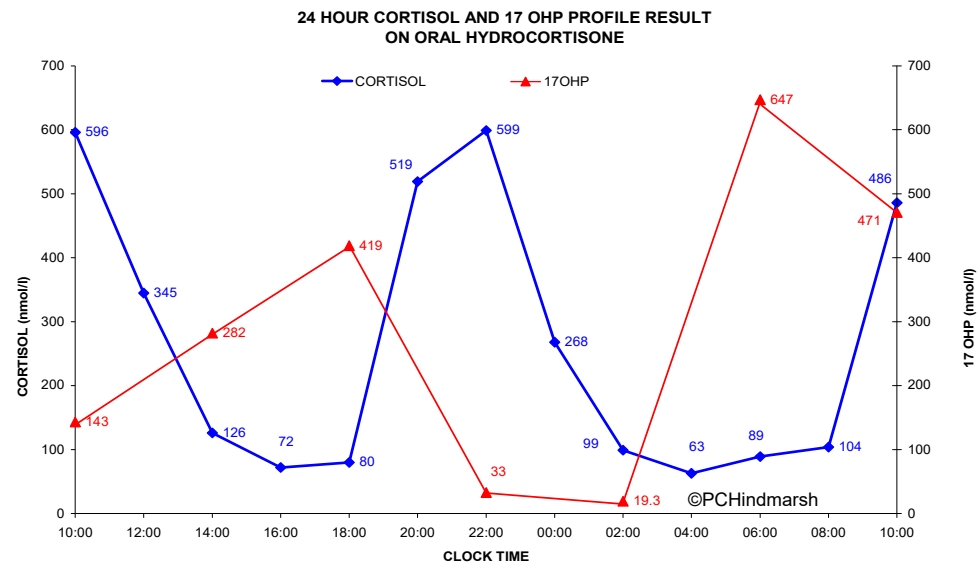
Pumps are becoming smaller and wireless pumps are now available on the market and a new slim, versatile, discreet rechargeable pump is due to be released soon. The inserters are so good it makes inserting the small cannula virtually painless. The site only needs to be changed every three to four days.

Bolus Function

There is also the option of using a bolus button, which infuses a bolus dose of hydrocortisone directly into the blood stream. The bolus can be given in various doses by the simple press of a button; some pumps have a remote control device.

Example One

The data in the graph below (Figure: 1) shows the result of a 24 hour profile done whilst Patient One was taking hydrocortisone tablets. This patient has a reasonably normal clearance rate of cortisol, however despite high peaks of cortisol, the 17 OHP remains high. We can see that the cortisol distribution over the 24 hour period does not follow the circadian rhythm.



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The data in graph below (Figure: 2) shows the result of a 24 hour profile done whilst the same patient, Patient One, is using the pump with the rates worked out using the Peter Hindmarsh Formula.

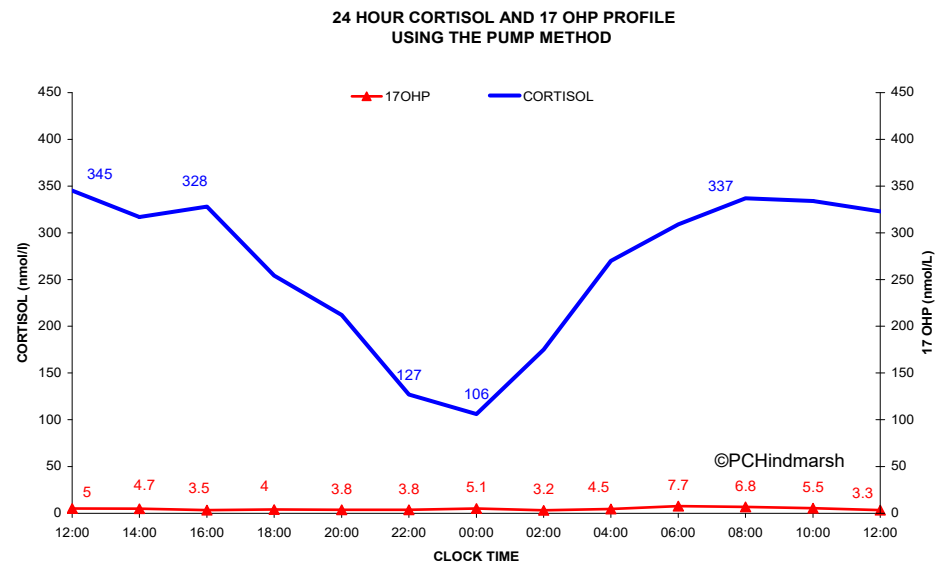


Figure: 2

The results in Figure 2 show us: -

1. With the right distribution of cortisol over the 24 hour period the 17OHP are stable and at the optimal level, similar to someone who does not have CAH.
2. The highest cortisol peak using the pump method is far less than that on the profile done when taking hydrocortisone orally, thus helping to prevent the side-effects that can be caused by over treatment. The highest cortisol level is occurring at the time of day when the body should naturally produce cortisol.

The cortisol delivered by the pump using the Peter Hindmarsh Formula is not only mimicking the circadian rhythm but we can see by the optimal suppression of the 17 OHP levels that the amount of cortisol delivered is being infused at the right rate, i.e. the right dose throughout the 24 hour period to suit this individual's metabolic needs.

This optimal control not only greatly improved the patient's general health and well-being, but side effects such as headaches and lethargy disappeared. This patient also suffered from severe gastritis which has now cleared up, as the pump delivery bypasses the gut.

Example Two

The data in the graph below (Figure: 3) shows the result of a 24 hour profile done whilst Patient Two was taking hydrocortisone tablets in four doses a day.

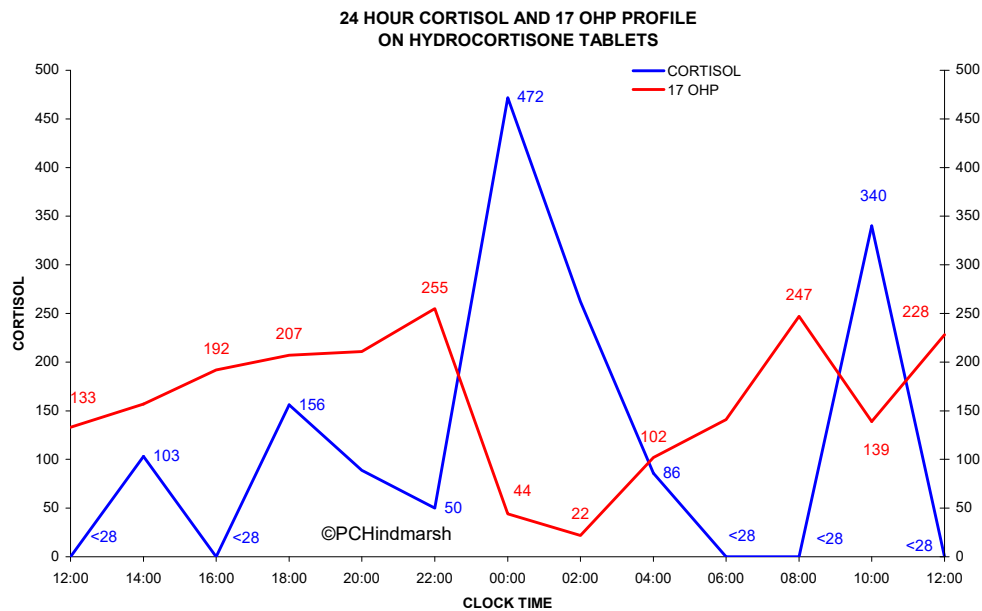


Figure: 3

The data in the graph above (Figure: 3) also shows that this patient clears cortisol rapidly. This is more common than we think in CAH and even though the patient was taking 4 doses of hydrocortisone a day, there are still periods where no cortisol can be measured in the blood and the 17 OHP remains high, despite this patient being on a high dose of steroid spread throughout the 24 hour period.

24 HOUR CORTISOL AND 17 OHP PROFILE ORAL HYDROCORTISONE 6 DOSES A DAY

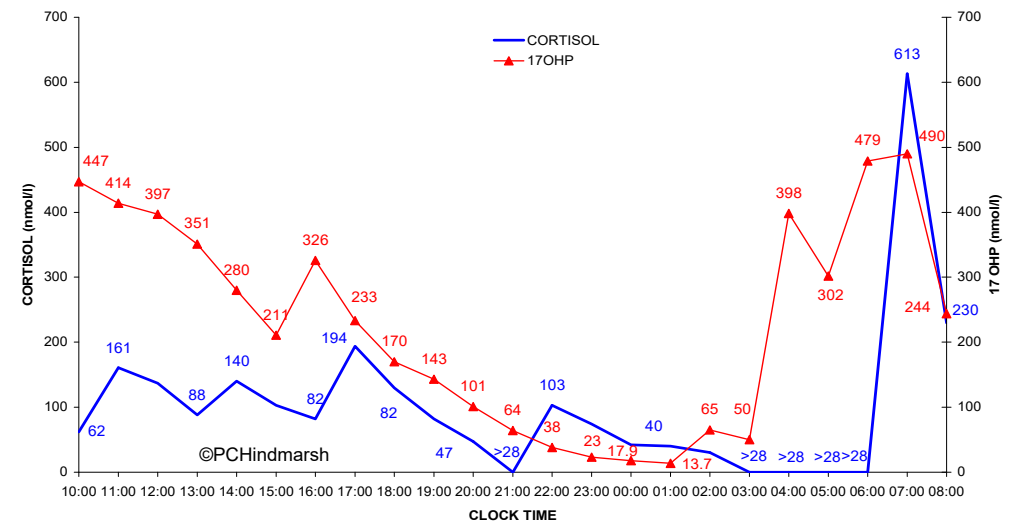


Figure: 4

The data in the graph above (Figure: 4) shows that despite Patient Two taking 6 doses of hydrocortisone a day, there are still periods where the patient is without any cortisol in the blood stream and despite a very high morning cortisol peak, the 17OHP levels are still high.

24 HOUR CORTISOL AND 17 OHP PROFILE USING THE PUMP METHOD

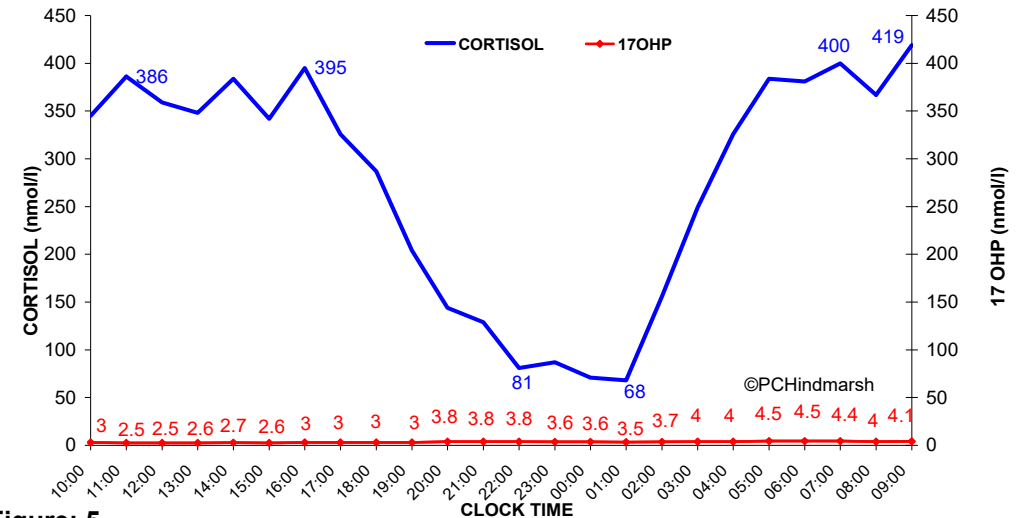


Figure: 5

The data in the graph above (Figure: 5) shows the result of a 24 hour profile done on the same patient (Patient Two) using the pump delivery method with the Peter Hindmarsh Formula. This can be compared with the data from the same patient show in Figure: 3 and Figure: 4.