

Successful Treatment of Addison's Disease with Continuous Subcutaneous Hydrocortisone via Insulin Pump



Jenkins MA¹, Crock PA^{1,2}, Nunn E¹, Bensing S³

1. John Hunter Children's Hospital, Newcastle NSW
2. Hunter Medical Research Institute (HMRI); GrowUpWell® PRC, University of Newcastle NSW
3. Karolinska Institute, Stockholm Sweden

Aim

To report a successful case of an insulin pump used to deliver continuous hydrocortisone for treatment of Addison's disease (AD) in an adolescent with inflammatory bowel disease.

Clinical features

The patient, a 17 year old female had a significant history of auto-immune disease. She was diagnosed with Addison's disease in 2015 after presenting to hospital with a life threatening adrenal crisis.

Other significant medical history included inflammatory bowel disease (IBD) and Hashimoto's thyroiditis (both auto-immune conditions), migraine and depression.

Her medications included

- Hydrocortisone physiological replacement therapy four times a day

4am	6mg
11am	6mg
3pm	4mg
8pm	4mg
- Fludrocortisone 200mcg daily
- Mesalazine 1.5g twice daily
- Topiramate 75 mg daily
- Venlafaxine 75 mg at night

The adrenal insufficiency caused by Addison's disease was treated with oral hydrocortisone.

However her inflammatory bowel disease was compromising absorption of the hydrocortisone, resulting in cortisol insufficiency. Increasing oral doses of hydrocortisone caused Cushingoid side effects.

She suffered lethargy and joint pain in her lower limbs. The joint pains (not due to inflammatory arthritis) were probably secondary to adrenal insufficiency. This limited her mobility and stamina and adversely affected her quality of life.

A trial of subcutaneous hydrocortisone was proposed, using an insulin pump to deliver continuous therapy to mimic natural cortisol production, with small boluses at breakfast and lunchtime.

Interventions, Case Progress and Outcomes

Concerns around using hydrocortisone via pump included

- "Off-label" use. Ethics advice was not considered necessary to trial the pump, which is already used for children requiring insulin. The IPU (Individual Patient Use) process was used instead through the hospital's drug and therapeutics committee.
- Infection control. Insulin contains preservative but hydrocortisone does not, so strict infection control procedures were necessary. As the infusion would be changed every 3 days, it was impractical to make a compounded product under aseptic conditions. Instead, Solucortef Act-o-Vials® were used to minimize manipulation at home and subsequent infection risk.

Practical aspects of giving hydrocortisone via pump:

- Pump rates were based on data from patients in Sweden using the protocol from the Karolinska hospital
- Pump rates varied throughout the day to mimic normal diurnal variation
- The patient and her parents were educated on the use of the pump, loading hydrocortisone and programming the dose
- A "sick day" plan was developed, and the patient instructed to increase basal rate to 200% in case of illness.

Monitoring

- The patient was admitted to hospital for 30 minute sampling of cortisol (plasma and saliva) and ACTH values to try and optimise pump rates.
- AddiQoL (Health-related Quality of Life in Addison's disease), a validated tool was used to show the change in her quality of life before and after starting the pump therapy (Figure 1). A total score from the sum of all the items is calculated, ranging from 30-120. A higher score is indicative of a higher quality of life.

Pre test score: 63

Post test score: 99

This indicates a significant improvement in the patient's quality of life, as a result of the hydrocortisone pump therapy.

AddiQoL						
Health-related Quality of Life in Addison's disease						
The following questions ask for your views about your health over the last 4 weeks and how you feel about life in general. Do not spend too much time answering, as your immediate response is likely to be the most accurate. Please answer every question.						
	None of the time	A little of the time	Some of the time	A good bit of the time	Most of the time	All of the time
I feel good about my health						
I can keep going during the day without feeling tired						
Normal daily activities make me tired						
I have to struggle to finish jobs						
I have to push myself to do things						
I lose track of what I want to say						
I sleep well						
I feel rested when I wake up in the morning						
I feel unwell first thing in the morning						

Figure 1. Example of AddiQoL questionnaire- tool for assessing quality of life

Discussion

This patient required an alternative to oral hydrocortisone for physiological replacement, as she did not have reliable oral absorption, and the pump has been very successful. She has responded well to the continuous infusion and has managed the practical aspects of pump therapy with minimal difficulty. She has not required hospital admission since starting the pump and has had minimal sick days.

There are clear advantages of delivering hydrocortisone delivery through a pump, including more closely mimicking basal cortisol levels.

However this solution may not be suitable for all patients. The need for self injection and wearing of an external device that may interfere with work and social activities, as well as serve as a constant reminder of their illness, may be unacceptable to other patients with Addison's Disease.

Conclusion

At 12 months the patient remains on subcutaneous hydrocortisone therapy and her improvement has been sustained. Her quality of life has dramatically improved, allowing her to study and work part time with no sick days.

For this patient, the successful use of hydrocortisone given via pump has been life-changing.

While there is evidence for continuous subcutaneous hydrocortisone via pump in adults, this case illustrates it can be used successfully in adolescents.

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