



Congenital Adrenal Hyperplasia Support Group of New Zealand

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Editorial

Summer arrived late for many in this country but what a glorious time it now is. Despite the delights of this warm weather, our CAHNZ Trust "business" continues much as usual. In this newsletter we offer a variety of up-to-date information with a focus on how members with CAH can maintain optimal health. Our support group receives regular enquiries, such as: "How do I remember my midday dose?", "My doctor is suggesting I change medication. How do I know if this is right for me?", "How can I best manage my weight?", and "I've just had surgery but have no guidelines about stress-doses of steroids. What shall I do?"

This Trust's role is to point members in the right direction to find answers to these questions. The starting place is almost always with their own physician. However such as those in rural areas or those newly-arrived to the country do not always know how to access some services. This is

where we can help and, if necessary, make suggestions about how seek a second opinion. Mostly, enquirers however simply want to talk to someone who 'understands' and is familiar with some of the complexities of having a rare condition like CAH. They want a sounding board as they work through their concerns.

A forum was presented recently at which members of CAHNZ were able to 'have a voice'. This was offered at a Round Table discussion in Auckland hosted by the Human Rights Commission. The catalyst for this forum was the collaborative work of the Human Rights Transgender Inquiry which resulted in the Jan 2008 policy document, "To Be Who I Am/Kia noho au ki toku ano ao". Intersex adults contributed to this document alongside those who identify as transgender. As a result several areas were identified that invited further dialogue for those who had

had previous genital surgery.

Two CAHNZ Trustees attended this event along with lawyers, adults with intersex conditions, doctors, researchers and nurse educators. It was a full and satisfying day to which the concerns of parents in this support group were well represented. A large number of questions arose, including the need for an audit of outcomes of genital surgery in more recent years for girls in Australasia and the need for recognition of the hardships many intersex adults have undergone as a result of prior or non-consensual medical treatment, exacerbated further by barriers presented by medico-legal bureaucracy. It was good to be part of this exploratory process. I hope we can continue to find ways of making progress in these very important areas.

Best wishes
CAHNZ Director



CAHNZ Newsletter #35 (Nov 2009):

"The article by the mum/midwife regarding breast-feeding (November 2009 CAHNZ newsletter) for painful procedures is topical. We routinely give neonates oral sucrose liquid before all painful procedures as there is a large research base proving that this results in less evidence of stress (both clinical pain response and endocrine markers). The same has been also shown for breast-feeding and breast milk given by dropper. Where there is a conflict between the needs of the medical staff for access, and the position of the baby for feeding, sucrose is an excellent option with the baby then going to the breast immediately after the procedure."

Dr Philip Moore, Paediatrician,

Hawkes Bay DHB, New Zealand

Welcome New Members

Jim & family

Sandra & family



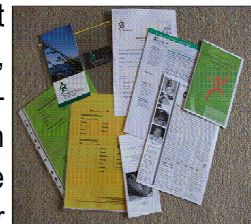
Hydrocortisone dosing during puberty in CAH (continued from page 3...)

- ◆ New Zealand also has the means for filter paper testing which means a close eye can be kept as needed on treatment doses related to growth acceleration. Thus growth and filter paper testing results can be individualised on a case by case basis.
- ◆ Drs Warne, Esko and Hofman also state that, in their experience, the dose quoted in the German survey (17mg/m²/day) is an unlikely treatment regime to be recommended for Australasian children. The usual dose range in treating Australasian children with CAH is approximately 10-13mg/m²/day. This is considerably less than the German study above.
- ◆ Finally, these doctors emphasise that the key is NOT in overdosing in puberty and that tight control of growth at this developmental phase is not as important as it is earlier in childhood.

**Visit our website:
www.cah.org.nz**

March Report

"The last four months have shown steady progress with support group work as Trustees have continued to work over the summer break on a number of projects, including the final touches to neonate nurse specialist Leigh McCarthy's Information Kit for CAH Newborns. These kits are now in the hands of neonatal nurses across the country and will shortly be sent out to paediatric departments as well. We invite any department or service who might want these kits to contact us. In addition, Leigh's information brochure for parents of newborn CAH babies has been printed and this much-needed resource is now freely available from CAHNZ or Leigh at 021 777 896.



The Trust continues with monthly teleconference meetings and is preparing for a third AGM on the 8th May 2010 for the first time in the South Island. A Christchurch venue has been booked at the Airport Gateway Motor Lodge Conference Room and *all members* are warmly invited to join Trustees for the AGM from 10.30 am - 11.30am and/or a lunch from 12.30—1.30 pm. Lunch costs will be subsidized but attendees will be required to contribute \$15/person. I will be emailing a draft agenda to members closer to the time.

It is very good to report steady progress on a number of CAH issues however we continue to need support and assistance on a number of projects and would particularly value help for newsletter research and production. If anyone has skills in this area, please contact the CAHNZ Director.

I hope to meet up with many of you at our May meeting."

Sandra

March 2010



Hydrocortisone Dosing During Puberty in C.A.H.



This is a review of an article published from the University Children's Hospital in Munich, Germany, recently published in full in the American CAH newsletter, and reviewed & condensed here by Dr. Heard, GP, Carterton, NZ.

The authors of this article presented their observations of the growth of ninety-two children with classical CAH whom they had treated exclusively with hydrocortisone (as cortisol replacement). Some received fludrocortisone if they were known to be salt wasting. These patients had had their growth pattern, final height & hydrocortisone (HC) dose monitored from the time of their birth until they had completed their growth at puberty. This group with CAH was comprised of all those treated at this centre and born between the years 1969-1987. The patients were monitored every three months during the first two years of their lives and then every six months during childhood and adolescence. All of the patients received HC dosing three times a day.

This German research is significant for a number of reasons:

- ◆ All ninety-two patients were treated at one centre over a twenty year period, thus reducing the variation in interpreting results (as opposed to comparing the growth of children treated in various centres).
- ◆ None of this group of patients received any other forms of treatment, e.g. gonadotrophin releasing hormones, which can be used to delay puberty.

The management of growth in children with CAH during puberty is difficult as a synthetic hormone, hydrocortisone (HC), is being substituted three times daily for a naturally occurring hormone (cortisol) which is continuously released in pulses throughout a 24-hour cycle from the adrenal glands.

Under treatment with HC leads to androgen excess which leads to advancement of bone age. This produces a reduced final height as the long bones of the body close off stopping any more upward growth. Overtreatment with HC also can cause suppressed growth by the growth inhibiting effects of glucocorticoids, of which HC is the one referred to here. Both under- and over-treatment with HC can result in a lesser height than one that is predicted from the parents' height. The rate of CAH children's growth as well as skeletal maturation

must always be closely monitored. A child's growth velocity is usually 5-6 cm per year from the time they turn 2 up until they start to go through puberty. Then growth can accelerate to 10-12cm per year.

The authors state that in their results they found that pubertal growth was significantly reduced in females with the average growth in puberty being only 13-14cm versus a reference population of 20.3cm. For pubertal males their results showed a pubertal total growth of 16-18cm versus a reference population of 28cm. This implies that the 'average' female with CAH would be 6-7cm shorter than what she would expect to be & a boy with CAH would be up to 12cm shorter than expected.

The results also show that during puberty the average daily HC dose was 17-18mg/m squared/day. They found a significant correlation between average daily dose of HC and final height. From their analysis they state that the upper daily limit of HC dosage during PUBERTY should be no more than 17mg/m squared/day.



The theory as to the cause of the decreased pubertal growth spurt are a 'too tight control' of growth with HC at the onset of puberty, which results in a decreased influence of sex hormones which normally would accelerate growth.

The authors state that pre-pubertal growth was not poor meaning that the discrepancy in final height is more or less due to a lesser pubertal growth spurt.

In summary, final height for these children with CAH was in the lower range of that predicted by using parental height as the indicator.

A NEW ZEALAND AND AUSTRALIAN PERSPECTIVE:



Three Australasian paediatric endocrinologists, Professor Gary Warne from Melbourne, Dr. Paul Hoffman from Auckland and Dr. Esko Wiltshire from Wellington, were asked to review this German research paper and compare with current

treatment regimes for Australasian CAH children. These doctors make some interesting points which must be taken into account when reading overseas research findings.

It is their consensus that Australasian children in more recent times have reached their expected final heights, compared to those in the German study whose final adult height was shorter than predicted.

Some reasons for this are as follows:

- ◆ The newborn screening with the Guthrie heel prick test means children with CAH (all severe and some milder forms) are diagnosed and treated generally before 2 weeks of age. (Note: Australia does not yet have newborn testing.)

(Continued on page 2...)

CAH in Males:

A Brief Overview

Dr Steven Soule

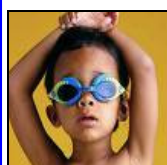
Endocrinologist, Christchurch Hospital, NZ

Email: steven.soule@cdhb.govt.nz



Infancy

It is no surprise, in view of the marked effects of the increased levels of adrenal androgens on female genital development, that there is a large amount of information available regarding the treatment of CAH in *females*. But there is surprisingly little information published regarding *males* with CAH. Indeed, in many parts of the less developed world, male CAH infants (particularly firstborn males) are not correctly diagnosed as having CAH and are merely thought to have gastroenteritis or an unexplained severe infection – without adequate steroid treatment many of these infants die. Of course the diagnosis of CAH in female infants is much easier to make as the genitals are obviously affected – this clue is missing in male babies in whom the genitals may look entirely healthy, although often there is darkening of the skin over the testes. In the developed world screening for CAH in the early neonatal period has greatly increased the detection of CAH, particularly in affected boys, and this has virtually eliminated the possibility of missing the diagnosis and not treating appropriately.



Childhood

Boys growing up with CAH walk the same treatment tightrope as their female counterparts – if they are *undertreated* with steroid then the adrenals will produce excessive amounts of androgens. This will stimulate the growth plates at the ends of the long bones and the boys will have an early growth spurt with an early and rapid puberty. The effect of this so-called 'precocious puberty' is that the ends of the long bones close off rapidly and the child ends up as a relatively short adult. The flip side of the coin is that if the boys are *overtreated* with ster-

oid (hydrocortisone plus or minus fludrocortisone) then although this will switch off the adrenal androgen production effectively, the steroid treatment itself may cause many side effects, including growth slowing and ultimately short stature. There are several points which help the Doctor decide on the best treatment through childhood – factors to consider include the growth rate, changes in body weight, levels of androgens (especially 17-OH progesterone, androstenedione and testosterone) as well as the rate of maturation of the skeleton, the 'bone age'. The practical message for parents and patients is that close follow up by a Paediatrician experienced in the management of this complex and relatively unusual condition is essential.



Adulthood

The adult male with CAH faces the usual challenges of taking the correct amount of glucocorticoid (usually hydrocortisone but sometimes prednisone or rarely dexamethasone) and often fludrocortisone to replace the deficiency of these hormones from the adrenal gland. As in childhood, there is a fine balance between *undertreatment* with the associated risk of tiredness and possible 'adrenal crisis' and *overtreatment* which carries with it a risk of weight gain, thinning of the skin, raised blood pressure and possibly even thinning of the bones. Once again, regular visits to an Adult Endocrinologist are important to monitor progress and to ensure that treatment is optimal – treatment that may suit a child or adolescent will probably not suit an adult so ongoing review is necessary.

Another interesting point is that there is evidence to suggest that the testes, the sperm and testosterone-producing factory in the male, may be abnormal in some CAH patients. There seem to be two potential reasons for damage to the testes. Firstly, as the male foetus develops in the womb the adrenal glands develop alongside the testes, and it is not at all uncommon for healthy males to have some adrenal tissue in their testes – this is called 'adrenal rest' tissue. In the CAH patient we know that the adrenal glands are driven to work extra hard by a hormone called ACTH (adrenocorticotrophic hormone)

from the pituitary gland. This means that the adrenal glands of patients with CAH may often be a little larger than normal. The same enlargement may occur in the adrenal rest tissue in the testes and this may cause small lumps in the testes and interfere with the functioning of the normal testicular tissue.

The second issue is that the high levels of adrenal androgens in CAH can switch off the function of the part of the pituitary gland which controls the testes with the result that the testes 'go to sleep'. The result may be a low sperm count, reduced fertility and sometimes low levels of testosterone in adult males with CAH. Fortunately there is evidence that these problems may be prevented if detected early, another reason for ongoing monitoring on a regular basis. The suggested approach is for all adult males with CAH who desire fertility to have a regular check by their endocrinologist on their testes along with possibly an ultrasound of the testes and sperm analysis to exclude any problems with the structure or function of the testes.

How common are these problems? Men with CAH less frequently have problems with fertility than affected females. Most affected males appear able to father children.

The practical message for parents & patients is that close follow up by a physician experienced in the management of this complex & relatively unusual condition is essential.

The limited available information on this issue suggests that 70% of adolescent and adult CAH males will

have an abnormal scan of the testes, 60% will have some abnormality of their sperm and about 30% will have a low level of testosterone – but it's important to realise that a low sperm count does not always preclude fertility. The good news is that if fertility is desired or if the testosterone level is low then changing the glucocorticoid treatment (hydrocortisone, prednisone or dexamethasone) may in some cases reduce the levels of the adrenal androgens and allow the testes to recover. However, in some patients removal of the adrenal rest tissue by an operation has been tried although it is still uncertain whether this is an effective way to improve the function of the testes. ■

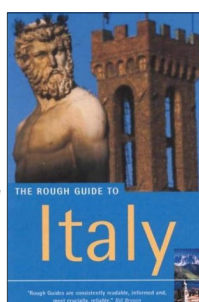
"Welcome to Holland"

"Welcome to Holland" is an essay written in 1987 by Emily Perl Kingsley about having a child with Down Syndrome though it is applicable to many other birth defects, and is given by many hospitals and child-care professionals to new parents of special-needs children.

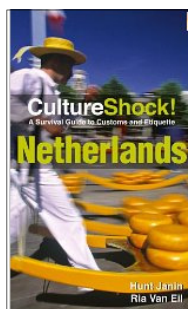


The essay, written in the second person uses a metaphor of excitement for a planned vacation to Italy that becomes a massive disappointment when the reader's plane lands instead in Holland...

"Holland?!?" you say. "What do you mean Holland?? I signed up for Italy! I'm supposed to be in Italy. All my life I've dreamed of going to Italy."



The metaphor is that the trip to Italy is a typical birth and child-raising experience, and that the trip to Holland is the experience of having and raising a special-needs child.



But everyone you know is busy coming and going from Italy... and they're all bragging about what a wonderful time they had there. And for the rest of your life, you will say "Yes, that's where I was supposed to go. That's what I had planned."

In the end, however, an effort is made to express that the "trip" is still well worth it:

But... if you spend your life mourning the fact that you didn't get to Italy, you may never be free to enjoy the very special, the very lovely things ... about Holland.

A song was also written in 2004 by Will Livingston based loosely on the story, also titled "Welcome to Holland".

Audio link for this can be found on the Broadjam site: www.broadjam.com

The full essay by Emily Perl Kingsley is as follows:

"I am often asked to describe the experience of raising a child with a disability - to try to help people who have not shared that unique experience to understand it, to imagine how it would feel. It's like this.....

When you're going to have a baby, it's like planning a fabulous vacation trip - to Italy. You buy a bunch of guide books and make your wonderful plans. The Coliseum. The Michelangelo David. The gondolas in Venice. You may learn some handy phrases in Italian. It's all very exciting.

After months of eager anticipation, the day finally arrives. You pack your bags and off you go. Several hours later, the plane lands. The stewardess comes in and says, "Welcome to Holland."

"Holland?!?" you say. "What do you mean Holland?? I signed up for Italy! I'm supposed to be in Italy. All my life I've dreamed of going to Italy."

But there's been a change in the flight plan. They've landed in Holland and there you must stay.

The important thing is that they haven't taken you to a horrible, disgusting, filthy place, full of pestilence, famine and disease. It's just a different place.

So you must go out and buy new guide books. And you must learn a whole new language. And you will meet a whole new group of people you would never have met.

It's just a different place. It's slower-paced than Italy, less flashy than Italy. But after you've been there for a while and you catch your breath, you look around.... and you begin to notice that Holland has windmills....and Holland has tulips. Holland even has Rembrandts.

But everyone you know is busy coming and going from Italy... and they're all bragging about what a wonderful time they had there. And for the rest of your life, you will say "Yes, that's where I was supposed to go. That's what I had planned."

And the pain of that will never, ever, ever go away... because the loss of that dream is a very, very significant loss.

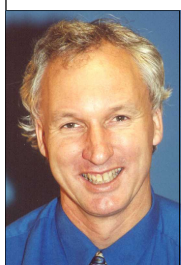
But... if you spend your life mourning the fact that you didn't get to Italy, you may never be free to enjoy the very special, the very lovely things ... about Holland."



Type 2 Diabetes Mellitus and Other Associated Adult Diseases in Women with CAH: Is there a link?

- Dr Paul Hofman

Paediatric Endocrinologist, Starship Children's Hospital



Diabetes mellitus encompasses a number of conditions that result in blood sugar (specifically glucose) levels being elevated. By far the most common is Type 1 diabetes mellitus (T1DM) which is much more frequent in children and Type 2 diabetes mellitus (T2DM)

which is the commonest cause of diabetes in adults. Although both result in high blood glucose levels they do so by very different mechanisms. In T1DM the problem is a profound deficiency of insulin. Insulin is the main hormone regulating glucose and increases the uptake of glucose from the blood into important tissues such as muscle, bone and fat. In T2DM the problem is not a lack of insulin but resistance to its actions. Thus to keep glucose levels normal in someone who has insulin resistance they have to secrete much more than usual. Over time the ability to secrete ever more insulin is reduced and T2DM ensues. While many factors regulate insulin action, the major factor in adults is the amount of body fat. Greater fat (especially on the trunk) is associated with greater insulin resistance and this is why most patients with T2DM are obese. More importantly insulin resistance is associated with a number of other adult diseases including high blood pressure, heart attack and stroke making the impact of insulin resistance greater than just the consequences of T2DM alone.

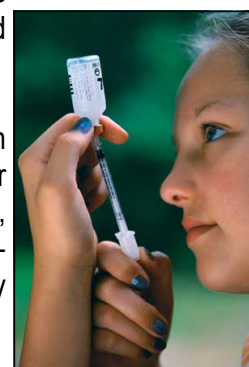
What has this got to do with CAH? CAH involves treatment with glucocorticoids at levels higher than those normally secreted by someone without CAH. These higher treatment doses suppress adrenal androgens which can masculinise women including excess male hair growth and acne. Treatment doses of hydrocortisone, prednisone, dexamethasone or other glucocorticoids can cause long term metabolic problems including greater body fat, osteoporosis (thin bones) and insulin resistance. Excess male hormone in women may also contribute to insulin resistance. *Does CAH increase your risk of T2DM and the other diseases associated with insulin resistance? The answer is it depends. If*

it increases weight and results in frank obesity then the answer is probably yes. In those who remain fit and lean with minimal evidence of excess male hormone the answer is almost certainly not. Are the risks of T2DM any different between those with or without CAH when amount of body fat is taken into account? The answer in women is a clear no. It is all about how fat or thin you are. The answer in men is unclear as there have been no studies in boys or men relating to long term metabolic changes and body fat accumulation.

Does CAH increase women's risk of T2DM and the other diseases associated with insulin resistance? The answer is it depends. If it increases weight and results in frank obesity then the answer is probably yes. In those who remain fit and lean with minimal evidence of excess male hormone the answer is almost certainly not.

In women there has been one excellent study from Sweden published in 2007 (J Clin Endocrinol Metab 92:110–116). Sixty-one women aged 18–63 yr, with CAH were compared to 61 age matched women without CAH (controls). Twenty-seven were younger than 30 yr, and 34 were 30 yr or older. Younger CAH patients and controls had similar amounts of body fat and insulin action. However older patients had higher body fat and insulin resistance compared to controls. Few older patients had high blood pressure, cardiovascular disease, or diabetes. Despite moderate glucocorticoid doses, most CAH patients had low androgens indicating their therapy was effective and possibly more than needed. Surprisingly diabetes during pregnancy was more common in CAH patients (21% of pregnancies vs. 0%). Diabetes during pregnancy often reflects impaired insulin action and is a risk marker for future diabetes. The study authors suggested life-long follow up with regular checks to ensure there are no adverse consequences of insulin resistance. This would include annual blood pressure assessment, and fasting blood tests to check lipids (blood fats) and glucose. Moreover the authors emphasised the need for long term lifestyle modifications, like diet and exercise, to improve insulin action as well as attempts to adjust and reduce the glucocorticoid doses especially in women older than 30 years. One weakness of the study is that glucocorticoid doses to treat CAH have changed over time and generally decreased. So older patients in this study would have been treated differently to the younger ones.

In summary, there may be an increased risk of T2DM and other insulin resistance diseases in CAH, especially closer to middle age. However the risk is low and seems mainly dependent on weight gain.



Magnesium

... An essential mineral for maintaining health & vitality!

"Vitamins B5, B6, Calcium & Magnesium are all required by the body to produce adrenal hormones.

(M. Woods, Naturopath, CAHNZ Aug 09)

Minerals such as zinc, iron and magnesium are essential nutrients that our body needs in small amounts for normal growth and development. Magnesium is one of the most important minerals—it is required for more than 300 biochemical reactions in the body! It helps maintain normal muscle and nerve function, helps our cells make energy and keeps our heart rhythm steady. Magnesium is also required for:

- Maintaining healthy blood sugar levels
- Breaking down protein, carbohydrate & fat to make energy
- Regulating calcium metabolism, helping keep bones strong
- Keeping our blood pressure low
- Improving blood flow around the body
- Enhancing muscle relaxation & relieving muscle cramps and spasms
- Helping us cope better with stress.

Are we missing magnesium?

Early signs of magnesium deficiency can include

- Tiredness, lethargy and fatigue—not having enough energy to get through the day
- Muscle cramping and spasms
- Muscle tension
- Tension headaches and migraines
- Stress, nervousness and anxiety
- Insomnia
- Premenstrual syndrome (PMS)
- High blood pressure.



For people experiencing one or more of these symptoms on a regular basis, or who have one of these conditions, they may be magnesium deficient.

Most diets are magnesium deficient

Magnesium is found in a wide range of foods, especially

green vegetables, grains, nuts, legumes and chocolate. However, the bad news is that it can be difficult to get an adequate supply of magnesium from our diet. In fact, a recent scientific study found that the daily intake of magnesium was below the Recommended Daily Intake (RDI) for 76% of men and 86% of women tested. Compounding this is that modern lifestyles actually increase our daily magnesium requirements significantly. For example, stress can deplete magnesium levels, as can a high consumption of tea, coffee and alcohol. Exercise can also increase our magnesium requirements.

Stressed out?

If we are stressed, we actually need more magnesium than usual to keep our muscles relaxed and keep our nervous system going. The irony is that during times of stress, we excrete more magnesium, leaving our stores of magnesium depleted. The depletion of magnesium can leave us feeling anxious, uptight and can even affect our sleep. This results in a vicious cycle, where stress lowers our magnesium levels and low magnesium levels increase our stress!

Break the stress cycle!

To break the vicious stress cycle, we may need to take some supplemental magnesium. There are many different forms of magnesium and having the correct form is extremely important when it comes to supplementation. Some forms of magnesium can cause gastric upsets and diarrhea for many individuals. Magnesium diglycinate is a form of magnesium that does not cause digestive upsets. This form of magnesium is highly absorbable and can help alleviate stress, anxiety and insomnia. Magnesium diglycinate is the best form of magnesium to take if you are magnesium deficient.



What's the best magnesium for me?

Magnesium diglycinate is available in an easy-to-take, pleasant-tasting powdered form from health stores. It also comes in combination with other important nutrients to help with symptoms of stress. For people with CAH, it is ideal to seek professional advice from a registered naturopath who understands the biochemical complexities of adrenal insufficiency and steroid replacement therapy, rather than rely on over-the-counter pharmacy advice or a pot-luck purchase off the supermarket shelf. Naturopath-recommended products are also available online, often at cheaper bulk prices. In addition to magnesium supplementation for people with CAH, practitioner Michael Woods (Herb Centre, Christchurch) also adds: "To combat some of the side effects of steroid treatment, supplementing with Vitamin B5 is essential."

For further advice contact: **The Herb Centre:** email clinic@theherbcentre.co.nz or phone (03) 3653 011.

a cool dry place is best

Regular reminders about medication storage are a regular feature in CAHNZ newsletters. This is because poorly-stored medication has reduced effectiveness over time, which can have significant long-term implications for the health of a person with CAH. Over the years members have from time to time reported how they or their children have developed unusual symptoms, with normally good routine blood tests beginning to give poor results. They are mystified at the shift in what has been for them a pattern of stable health. On occasion, doctors have challenged them about non-compliance in taking medication at the prescribed times or in the right dosages. In three quite recent accounts, the problem lay with not with non-compliance but with poor storage of medication. Sometimes this was the fault of the owner; others that of the provider. Examples are: (i) medication being couriered long-distance in a non-refrigerated truck; (ii) pills being stored in the kitchen above the oven; (iii) a new pharmacist preparing hydrocortisone syrup (the family were not informed).

People with rare and complex health conditions like CAH are required to manage their health very pro-actively on a day-to-day basis. This takes time, thought and commitment, and sometimes (being human) the importance of careful medication storage is overlooked. It is easy to sometimes leave lids off pill containers, keep pills in cars where they get sun-baked, or in pockets for a quick midday dose—where they get discovered a few days later and quickly popped without a second thought. Here are some further reminders:

1. Most medicines can be kept at room temperature. However some require either refrigeration or special storage conditions (e.g. Florinef). Silicon sachets can be requested at no charge from the pharmacist (or saved from other bottles, e.g. vitamins) and are useful for absorbing moisture in pill containers.
2. Do not keep medicines in the fridge unless the label says so; this may destroy their effectiveness.
3. It is best to store medicines outside of the bathroom, as steam and humidity from showers and baths may reduce the effectiveness of some drugs. If your pills are moist and powdery, that's an indication they've been affected by humidity and/or changing temperatures. Show them to your pharmacist before using them and ask about the best place to store them.
4. Check spare supplies regularly, both cortisol, Florinef and SoluCortef. Put a reminder in your diary or mobile phone to prompt you to do this every 3-6 months.



Remembering Your Pills

The Pill Box Timer is a handy gadget for those who have difficulty remembering their daily doses. (That midday one is often a pest!)



- Holds pills, tablets and capsules
- 24 hour timer
(Alarm will sound when set time is up.)
- Has two compartments for each measuring:
- Length: 5.5 cm, width: 2 cm, depth: 1.5 cm.
- Audible beep reminds you to take medicine on time
- Makes a short beep 5 or 10 minutes before set time.
- Price: NZ\$19 (incl. GST) plus P+P

Order online @ <http://www.disabilityresource.org.nz>

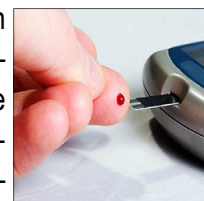
High Magnesium and Calcium Intake Linked to Lower Diabetes Risk

It is well known that diet plays an important role in the development of type 2 diabetes, but less is known about the influence of specific nutrients on non-Western populations. A report published in the March 2009 issue of the American Journal of Clinical Nutrition revealed a protective effect of calcium and magnesium against diabetes in a large group of Chinese women.

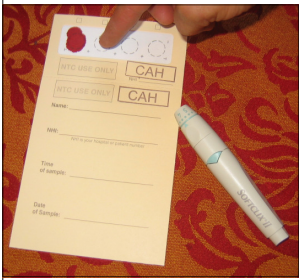
The study involved 64,191 women from the Shanghai Women's Health Study. All women lived in Shanghai, China. Analysis of dietary questionnaire responses determined calcium and magnesium intake.

Women whose intake was the highest at an average of 649.6 milligrams per day had a 27% lower risk of diabetes than those whose intake was in the lowest group at 277.5 milligrams. Women whose intake of magnesium was highest at an average of 318.1 milligrams per day experienced a 20% lower risk compared with those in the lowest category of intake. Dairy intake was also related to a lower risk of type 2 diabetes.

The researchers did not have information on vitamin D intake, but the protective effect of dairy products could be partly due to their vitamin D content as well as calcium. The combination of vitamin D and calcium has been associated with a reduction in the risk of type 2 diabetes in previous research. (Am J Clin Nutr 89:1059-1067, 2009)



and CAH: A 2010 Update



- Sheryl Tregurtha

Paediatric Diabetes and Endocrine
Nurse Specialist, Green-
lane Clinical Centre, Auckland,
New Zealand

Hi, I thought I would take this opportunity to introduce myself. I am the Paediatric Endocrine Nurse Specialist at Starship Hospital. Since the change in Christine McMahon's role, I am now the liaison nurse for CAH – 17 OHP Filter paper testing. This service will have a slightly different look as unlike Christine I am Auckland-based and unable to travel around the country.

However since taking over this role I have compiled an up-to-date brochure with simple step-by-step instructions on how to do the filter paper testing. In conjunction with Gabrielle McCarthy I have also put together an information pack for families with newly diagnosed children. This will hopefully ensure that wherever in New Zealand families are diagnosed they will have access to the same information.

I would encourage everyone with a child affected with CAH to regularly do a 17 OHP 24 hour profile. This means four to six finger prick tests / day. A general guide as to frequency might be 3 – 4 times a year, or as otherwise directed by your Doctor. This enables your Doctor to get a much clearer idea whether you or your child is on the correct medication, and if not, which dose needs to be adjusted.

If you are not currently doing this filter-paper testing, and would like more information then talk either to your doctor (paediatrician, endocrinologist) or their nurse. Alternatively you can contact me at the following email address:

diabnurse@adhb.govt.nz and "Attn: Sheryl"

or ring me on 09 631 0790 (option 2) – and leave a message. Please note however that I only work on Monday and Wednesday, so it may take a few days before I can get back to you.

Notice to All Members

CAHNZ Trust Annual General Meeting

8th May 2010

Venue: Airport Gateway Motor Lodge

Conference Room

@ 45 Roydvale Ave, Burnside, Christchurch

Time: AGM: 10.30 - 11.30 am

Lunch: 11.30-12.30 pm

Cost for lunch \$15

Please note: we would love to see you there ...



however - **RSVP is essential**. Please contact us at:
CAHNZ@snap.net.nz

For an online north-west Christchurch map, visit Wis-
es Maps @ [http://www.wises.co.nz//Christchurch/
Burnside/45+Roydvale+Avenue/#c/-
.513327/172.597275/13/](http://www.wises.co.nz//Christchurch/Burnside/45+Roydvale+Avenue/#c/-513327/172.597275/13/)

Information Booklet for Adults with CAH

Much historical emphasis on the care and treatment of people with CAH has been on children's growth and development. Until more recently, less information has been available regarding

adult well-being and fertility. In 2000, the 'Information Booklet for Adults with CAH' was written by UK Drs Conway and Stanhope 2000 and still provides a very good overview of CAH adult

health issues. A copy of this document can be downloaded in pdf format from www.ahn.org.uk along with other links for information and support.



Travelling with Medication:

Prescription for Trouble

DAUNTING: Getting through customs with prescription drugs can prove scary if you're caught unawares. Taking your medication with you should be hassle-free, if you follow the rules, writes **Jane E. Fraser**, Reuters.



International

It is not a good feeling, standing in front of a stern-faced customs official who has just found a bag of unidentified pills in your luggage. As a backpacker trying to lighten the load, I had the "clever" idea of emptying a bottle of multivitamins into a snap-lock bag to save taking the heavy glass bottle with me.

It turned out to be a rather big, and rather scary, mistake.

I found myself surrounded by armed personnel in the airport in Bangkok – and there are few places where drugs are treated more seriously.

They eventually let me go after I convinced them the pills were only vitamins but the experience was enough to ensure I never did it again.

Travelling overseas with medication, prescription or otherwise, can be tricky and requires a bit of forethought and planning.

How do you get hold of your regular medications if you're away for a long time? How do you know if you're allowed to take a certain medication into a country? And what else do you need to take with you to avoid a run-in with the authorities?

The first thing you should do if you are planning an overseas trip and need to carry medications – particularly strong painkillers or other drugs open to abuse or sale on the black market – is to contact the embassy or consulate for the country or countries you are visiting to make sure the medication is legal in that country.

Certain classes of prescription medicines are regarded as illegal substances in some countries and the consequences of carrying them can be dire.

Assuming your medications are legal to take, you should visit your doctor and, if possible, get a prescription to cover the entire time you are going to be away.

Fill the prescription before you leave – a prescription from your home country has no validity in other countries.

If you are unable to carry enough medication for your entire trip, make sure you take a letter from your doctor detailing the medication, dosage and what it is for, especially if it is a painkiller or another drug a doctor may be wary of prescribing.

A letter from your doctor explaining that your drugs are

medically prescribed and are for personal use can also ease your way through customs checkpoints.

Likewise, if you need to carry needles and syringes (it is best to take your own unless you are travelling to a developed country), a letter from your doctor is vital.

And, as the Bangkok scenario demonstrates, all medications and vitamins should be kept in their original packaging (plastic bottles are now available in most cases).

Medications should also be carried in your hand luggage in case your luggage is lost—and let the airline know in advance if you will be carrying syringes!

It is also a good idea to keep a doctor's letter or list of medications and dosages separate to the actual drugs, in case you lose your bag in your travels.

One issue travelers struggle with is medications that need to be kept refrigerated. Some airlines will put them in the fridge during the flight but others won't let you do it because of hygiene reasons. You need to check with the carrier in advance.

If you need to keep medications cold during your flight, gel-based ice packs stay cold for a long time, or you can take some "Snap Lock"® bags and ask the flight attendants for ice.

If you choose to take ice packs, they should be exempt from rules about carrying liquids on flights but make sure you declare them as you go through the screening point.

Once at your destination, a small insulated bag with gel-based cold packs is the best option if you have access to a freezer each night.

You can buy purpose-made medicine cooler bags at travel stores and over the internet. These are good if you need to carry a range of vials or containers or have very temperature-sensitive medicine.

Finally, make sure that you declare any drugs that you are bringing back to Australia, as certain classes of drugs are subject to import restrictions.

Counter-culture

You might be surprised to find the medication you get on prescription at home is readily available over the counter in other countries, although it could have another brand name.

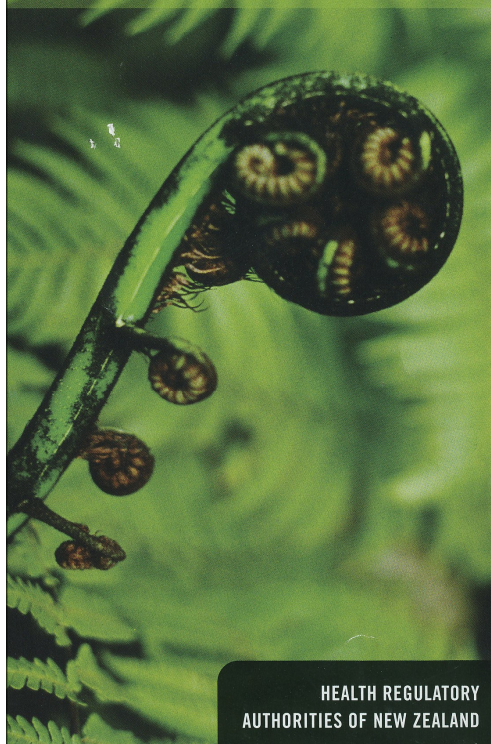
Unfortunately, the situation can also be reversed — something you normally buy over the counter at the chemist may require a prescription.

If you are buying any medication overseas, check the dosage — it may be more or less than the amount you normally take.



(Reference: www.stuff.co.nz)

Confidence in your health practitioner



HEALTH REGULATORY
AUTHORITIES OF NEW ZEALAND

What if something goes wrong?

The independent Health and Disability Commissioner promotes and protects the rights of consumers of health services and receives all complaints against health practitioners. If you have a complaint about a health practitioner you should contact the Commissioner's office on 0800 11 22 33 or www.hdc.org.nz.

The Act creates a separate Health Practitioners Disciplinary Tribunal. The Tribunal considers serious disciplinary matters and can impose penalties ranging from fining the practitioner, ordering that the practitioner practise only under certain conditions (for example under supervision), to suspension or cancellation of registration.

You may also want to talk to the practitioner's registering authority about your concerns. Web addresses and phone numbers for each authority are listed on the back of this pamphlet.

The Health Regulatory Authorities of New Zealand (HRANZ) is an informal group made up of the 15 registering authorities listed overleaf. HRANZ oversees and manages matters of common interest.

This leaflet has been produced by HRANZ with the support of the Ministry of Health.



www.moh.govt.nz

HP3995

REGISTERING AUTHORITY CONTACT DETAILS

Chiropractic Board	Ph 04 474 0703
www.chiropracticboard.org.nz	
Dental Council	Ph 04 499 4820
www.dcnz.org.nz	
Dietitians Board	Ph 04 474 0746
www.dietitiansboard.org.nz	
Medical Council of New Zealand	Ph 04 384 7635
www.mcnz.org.nz	
Medical Laboratory Science Board	Ph 04 474 0701
www.mlsboard.org.nz	
Medical Radiation Technologists Board	Ph 04 474 0745
www.mrtboard.org.nz	
Midwifery Council	Ph 04 474 0740
www.midwiferycouncil.org.nz	
Nursing Council of New Zealand	Ph 04 802 0247
www.nursingcouncil.org.nz	
Occupational Therapy Board	Ph 04 474 0708
www.otboard.org.nz	
Osteopathic Council	Ph 04 474 0747
www.osteopathiccouncil.org.nz	
Optometrists and Dispensing Opticians Board	Ph 04 474 0705
www.optometristsboard.org.nz	
www.dispensingopticiansboard.org.nz	
Pharmacy Council	Ph 04 495 0330
www.pharmacycouncil.org.nz	
Physiotherapy Board	Ph 04 471 2610
www.physioboard.org.nz	
Podiatrists Board	Ph 04 499 7979
www.podiatristsboard.org.nz	
Psychologists Board	Ph 04 474 0744
www.psychologistsboard.org.nz	

SEPTEMBER 2004

Confidence in your health practitioner

The main purpose of the Health Practitioners Competence Assurance Act 2003 is to protect the health and safety of the public. It seeks to ensure that the treatment and healthcare you receive from a registered health practitioner is of a high standard.

This leaflet tells you how the Act benefits you.

The Act covers all registered health practitioners and replaces a number of separate, outdated laws. The Act looks after your health and safety in the following ways:

- It ensures health practitioners are properly trained and qualified before they can be registered
- It requires health practitioners to continually update and improve their skills
- It establishes independent registering authorities to register and monitor health practitioners
- It establishes an independent Health Practitioners Disciplinary Tribunal.

What does the Act cover?

The Act covers all registered health practitioners in New Zealand, including:

- chiropractors
- dentists, dental technicians, clinical dental technicians, dental therapists and dental hygienists
- dietitians
- dispensing opticians
- medical laboratory scientists and technicians
- medical practitioners (such as doctors, psychiatrists, surgeons and other specialists)
- medical radiation technologists
- midwives
- nurses
- occupational therapists
- optometrists
- osteopaths
- pharmacists
- physiotherapists
- podiatrists
- psychologists.

In the future, other health practitioners may also come under the Act.

How are you protected?

Under the Act, each practitioner is answerable to an independent registering authority (listed over).

Registering authorities ensure that health practitioners:

- are registered in a 'scope of practice' which describes the health services they can provide
- have the right qualifications for their scope of practice
- have a current practising certificate (license) which has to be renewed each year
- maintain and develop their skills and competence
- have good English language and communication skills
- are physically and mentally able to work.

Each registering authority maintains a register or list of health practitioners that is available to the public. In many cases you can find and look at a register on the Internet.

Monitoring health practitioners

Registering authorities set the standards that all health practitioners must meet. If a registering authority has concerns over the quality and standard of treatment a practitioner provides, it may take steps to protect the public. For example, it may require a practitioner to undertake some further training, practise under the supervision of another practitioner or limit the kind of services he or she can provide.

Question:

"Our daughter with CAH had a check up with a new paediatrician at four years of age. It came out by chance that she was still taking hydrocortisone syrup. We had been getting some repeats over the phone and the issue just had not come up in other consultations. The new paediatrician was aghast and promptly changed her onto pills. This made life much easier for us in terms of storage and also stress-dosing during illness. We felt silly as we should have brought this up earlier, but visits were always so busy and there was always lots to talk about. Can you tell us, what are the guidelines for parents of CAH about when to shift their child from syrup to pills? If a very young child can take pills, can syrup be replaced pretty early on?"



Answer:

Wellington paediatric endocrinologist, Dr Esko Wiltshire, replies:

"There is no 'set time' when a change over from syrup/elixir to pills should occur for children taking hydrocortisone – it comes down to convenience and being able to give a suitable dose. Hydrocortisone elixir is made either from powder or sometimes from the tablets. It has a very short shelf life (7-10 days), after which it becomes unreliable, unless it is made up with a particular preservative, which is both expensive and difficult to obtain. This really means collecting the elixir from the pharmacy on a weekly basis, as you all know! There is no problem with using the liquid for as long as you like, as long as it is collected fresh each week. The tablets avoid this problem, as they have a long shelf life, which also allows for easier access to stress dosing when needed. In New Zealand we can only get 5 mg tablets, which can be divided into quarters (1.25 mg), although not always easily. I usually swap infants to tablets when we are close to doses that are multiples of 1.25 mg (e.g. a half tablet in the morning, quarter in the afternoon and quarter at night). There are 1 mg hydrocortisone tablets available overseas, but thus far it has not been possible to get these in New Zealand. This is usually sometime between ages 1 and 2, but will vary from one child to the next. Fludrocortisone can be given as a tablet (crushed or dissolved) from birth, as infants are usually on doses that can easily be given as a whole or half tablet."

Emergency Department Doctor

Question:

"I understand a person with congenital adrenal hyperplasia has a form of 'adrenal insufficiency' and in the case of serious illness or accident may have an adrenal crisis or 'collapse'. How serious is this, and how should one put plans in place to reduce the chance of crisis?"



Answer:

"Anyone who does not produce the stress-hormone cortisol is at risk of adrenal crisis if they do not get prompt help with the following: blood loss, serious injury, fluid and electrolyte loss (e.g. vomiting, dehydration), infection (including fever), severe diarrhoea. It doesn't matter if you have Addison's or CAH, the issue is the same. A state of adrenal crisis is considered a **medical emergency!** There are a few preventative as well as treatment options to keep in mind:

1. All people with CAH should have an 'action plan'. The basics of such a plan are:

- (i) Always wearing a Medic Alert disc that states "Adrenal Insufficiency"; taking steroids;
- (ii) Keeping an easy-to-locate and up-to-date emergency kit with injectable cortisol (SoluCortef) PLUS needles PLUS a copy of a doctor's letter advising emergency guidelines;
- (iii) Informing family/friends/partner/flatmates where you keep your emergency kit, and keep your physician's telephone number and emergency medical phone numbers handy. It's ideal if someone other than yourself (if you are an adult) can give an intramuscular injection *before* the ambulance arrives or *before* you go to an emergency department.

2. Proper emergency medical response to a person with CAH will include the following treatments:

I.V. or I.M. cortisol injection (SoluCortef), along with isotonic saline. I.M. or I.V. cortisol should be continued until oral medications can be tolerated. I.V. glucose should also be administered in the case of hypoglycaemia.

