



11 April 2008

Lord Ara Darzi  
**Our NHS, our future** review  
Department of Health  
*via email*

Dear Lord Darzi,

## **I. Overview: current initiatives within the NHS**

National working groups are currently considering how best to embed goals of quality improvement, innovation, effective primary care, workforce skills and leadership development within the NHS. At the same time, local working parties within each Strategic Health Area are considering how to plan for the health care needs of the community across the spectrum from birth to old age, taking account of both chronic, long-term conditions and acute medical needs.

We wish to bring to your attention the needs of a small group of patients whose needs straddle the remit of both national and local working groups. These patients have a rare, potentially-fatal endocrine condition that requires complex daily medication, regular, long-term medical care and occasional acute treatment: Addison's disease. Best estimates indicate that there are less than 8,800 cases of diagnosed Addison's across the UK, of whom up to 700 per annum can be anticipated to need acute, emergency treatment to prevent a potentially-fatal adrenal crisis.<sup>1</sup>

## **II. Core recommendations**

We wish to suggest that health outcomes for this group of patients would be enhanced by establishing:

- Regional (tertiary) centres of expertise in adrenal medicine
- Effective partnerships between tertiary centres and local GPs.

## **III. Addison's disease: acute and chronic medical dependencies**

People with well-managed Addison's disease can lead long and productive lives. It is not unknown for people with Addison's to live into their nineties, to raise a family as a single parent, to complete marathons or triathlons, or to hold down demanding jobs. One member of the ADSHG was recently awarded an MBE for military services, some 18 months after his diagnosis<sup>2</sup>.

Conversely, undiagnosed or under-treated Addison's disease can rapidly lead to death or permanent disability, usually resulting from hypoxia or cardiac complications. A few members of the ADSHG have been left unable to work through physical or intellectual disability as a result of delayed treatment for adrenal crisis. Our 2003 membership

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<sup>1</sup> See <http://www.endocrine-abstracts.org/ea/0013/ea0013p113.htm> for the causes and frequency of adrenal emergencies

<sup>2</sup> See attachment 1



survey found that 10% of respondents were unable to work through disability, whereas some respondents were able to work 60 or more hours a week.<sup>3</sup>

Thus, good medical care and the right balance of daily medication are essential for people with Addison's to lead full and active lives and to participate in the workforce. This requires:

- 1. Good medical support from an informed GP:**
  - including adequate and timely supplies of essential prescription medications.
- 2. Ongoing medical monitoring from an adrenal specialist (tertiary endocrine service) to:**
  - ensure the medication balance remains optimised
  - monitor for the likely development of associated autoimmune conditions.
- 3. Prompt emergency treatment for adrenal crisis, requiring:**
  - a coordinated response from the after-hours GP service, ambulance service and Accident & Emergency staff.
- 4. Adequate adjustment/supplementation of steroid medication for:**
  - physical challenges such as surgery, dentistry, pregnancy, cardiac disease or old age.

#### **IV. Role of the GP**

Because Addison's disease is a rare condition, few GPs can expect to see more than one such patient in their professional career. Therefore, the GP needs support and guidance from an adrenal specialist to ensure they:

- Understand how and when to adjust the normal replacement steroid medication for day to day requirements such as strenuous exercise, overseas travel.
- Understand how to adjust glucocorticoid medication for injury, fever, gastric infection or any infection requiring antibiotic treatment<sup>4</sup>.
- Appreciate the risks of adrenal crisis in cases of dehydration, shock or under-medication, and their role in liaising with emergency services to ensure prompt treatment.
- Can identify when to involve the adrenal specialist or other consultants as and when the patient shows signs of further health complications.
- Know when to consult the adrenal specialist regarding other modifications to the normal steroid replacement therapy, eg surgery, major dental procedures.

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<sup>3</sup> See <http://www.addisons.org.uk/comms/meetings/harrogateposter2005.pdf> for workforce participation rates

<sup>4</sup> Although many GPs will have a degree of familiarity with adjusting glucocorticoid therapy; none are experienced with mineralocorticoid medication. Mineralocorticoid monitoring must be conducted at a hospital laboratory with facilities to process plasma renin within 10 minutes of the sample being drawn.



In 2003, the ADSHG conducted a membership survey where we asked patients to rate their GP's level of knowledge about Addison's. Around the UK, 468 Addison's patients replied, which amounts to over 5% of the total UK patient population.

More than 40% felt their GP knew very little, or didn't know much about Addison's. Only 9% felt that their GP knew a great deal about Addison's.

We can, therefore, conclude that most GP's are currently under-supported in their role as primary caregiver for this group of patients.

## **V. The role of the adrenal specialist (endocrinologist)**

The rarity of Addison's means that many endocrinologists at smaller hospitals will only treat one or two patients, so that they will not be able to develop any depth of experience in the best management of the patient.

Thus, unsurprisingly, standards of endocrine care vary widely across the UK. Many members of the ADSHG report hospital practices that fall short of the recommendations of the ADSHG's Clinical Advisory Panel. Only those attending the largest teaching hospitals report standards of endocrine care that are fully consistent with our panel's recommendations for:

1. Optimisation of steroid replacement therapy by monitoring:
  - Mineralcorticoid replacement (plasma renin, serum electrolytes)
  - Glucocorticoid replacement (hydrocortisone day curve)
  - DHEA replacement (DHEA-S, testosterone and oestrogen)
  - Calcium and other indicators of bone turnover.
2. Life-long monitoring for the anticipated development of associated conditions:
  - Frequently, hypothyroidism or Graves disease, diabetes, vitamin B12 deficiency, coeliac disease or other autoimmune conditions<sup>5</sup>.
3. Patient education in prevention and treatment of adrenal emergencies, including training in self-injection.
4. Guidance to the patient's other medical specialists regarding steroid cover for surgical and dental procedure.

## **VII. Recent initiatives in disseminating best practice**

Discrepancies in the management of Addison's patients have prompted the Addison's Clinical Advisory Panel to develop a range of patient information materials<sup>6</sup> covering:

1. Treatment of adrenal crisis
2. Glucocorticoid cover for surgery and dentistry
3. Information for the newly-diagnosed patient
4. Information for the GP.

<sup>5</sup> See <http://www.endocrine-abstracts.org/ea/0011/ea0011p188.htm> for the incidence of associated conditions

<sup>6</sup> See <http://www.addisons.org.uk/publications> for PDFs of these publications



Several years ago, the ADSHG established an electronic discussion group (forum) and we have observed that our members now explicitly discuss the variability in standards of treatment at hospitals around the UK. More recently, patient choice has been utilized by some of our members to request that their GP refer them to the larger endocrine units with a greater depth of expertise in adrenal management. In particular, our members are increasingly motivated to seek treatment at a centre that can offer:

- Training in self-injection and the provision of injection materials
- Day curve monitoring of hydrocortisone replacement therapy
- Access to endocrine specialist nurse support.

### **VIII. Conclusion**

In light of the work of the Darzi review, it seems timely to reappraise the configuration of care that is currently shared between GPs and endocrinologists, with a view to encouraging regional (tertiary) centres of expertise in the management of adrenal insufficiency. These tertiary centres will need to work in partnership with local GPs, who remain responsible for day-to-day care and emergency medical support.

We would be pleased to offer further information and assistance on this matter, should you wish to discuss these issues in more detail.

Yours sincerely,

**Katherine G White**

**ADSHG Chair & Clinical Advisory Panel coordinator  
on behalf of the ADSHG trustees**

### **Attachments:**

