A case of hypertension and hyperkalaemia

Atef B Michael, Peter Wallis

INTRODUCTION

Gordon's syndrome is a rare hereditary metabolic disorder. It is characterized by hypertension, hyperkalaemia and hyperchloraemic acidosis. Renal and adrenal functions are normal. The biochemical abnormalities are reversed with thiazide therapy.

DISCUSSION

Gordon's syndrome or familial hyperkalaemic hypertension is also referred to as pseudohypoaldosteronism type II (PHA2). The syndrome is characterized by hypertension and hyperkalaemia, with normal kidney function and correction of the biochemical abnormalities by thiazide diuretics.

The pathophysiology of the disease is still controversial. Several mechanisms have been postulated. Gordon (1986) proposed that increased renal sodium absorption proximal to the aldosterone-regulated segments leads to decreased sodium delivery to the cortical collecting ducts, and hence decreased luminal negativity. The negative lumen potential helps potassium ions to diffuse across the tubular cell membrane.

Biochemically, the syndrome is characterized by hyperkalaemic, hyperchloraemic normal anion gap metabolic acidosis. The renin and aldosterone levels are low. Kidney and adrenal functions are normal.

The mode of inheritance in the reported families is consistent with autosomal dominant transmission. Three loci have been described for Gordon's syndrome on chromosome 1 (PHA2A), 17 (PHA2B), and 12 (PHA2C). Studies have demonstrated genetic heterogeneity and a fourth gene for familial hyperkalaemic hypertension (Disse-Nicodeme et al, 2001). The reports of linkage for familial essential hypertension within the same area of chromosome 17 containing the genetic marker of Gordon's syndrome have raised the possibility that the same gene might be involved in both conditions (O'Shaughnessy et al, 1998).

CASE REPORT

A 52-year-old man was referred urgently to hospital by his GP because he had hyperkalaemia (7.3 mmol/litre; normal range 3.5–5.0 mmol/litre). Two months previously he was diagnosed as having diabetes mellitus when he was admitted to hospital with diabetic ketoacidosis. At that time his plasma sodium was 126 mmol/litre (normal range 133–147 mmol/litre), potassium 2.7 mmol/litre, urea 18 mmol/litre (normal range 2.5–7.5 mmol/litre) and creatinine 159 μmmol/litre (normal range 60–120 μmol/litre). Bendrofluazide, nifedipine and atenolol were stopped. Six weeks later he felt unwell, visited his doctor, and was found to be hypertensive. He was started on lisinopril 5 mg once daily. Ten days later he was still hypertensive, and lisinopril was increased to 10 mg once daily and routine plasma urea and electrolytes were measured.

His past medical history includes a Colles' fracture in 1992. At this time, hypertension and hyperkalaemia were noted and investigated. He was diagnosed as having Gordon's syndrome. Three of his daughters and two of his granddaughters were also found to have the syndrome. His father had been hypertensive and died suddenly at the age of 40 years. His uncle had also died suddenly at the age of 33 years.

On examination his height was 1.60 m, body mass index (BMI) was 30 kg/m^2 and blood pressure was 168/75 mmHg. Heart, chest, abdominal, and neurological examination were normal. The electrocardiogram did not show manifestations of hyperkalaemia.

The hyperkalaemia was treated with insulin/glucose infusion. Lisinopril was stopped and bendrofluazide was started with normalization of plasma potassium level and blood pressure.

The age at diagnosis may be as young as 2 weeks or as old as the sixth decade. Achard et al (2001) studied 14 families with familial hyperkalaemic hypertension, and found that several subjects were asymptomatic. Many subjects have normal blood pressure at the time of diagnosis. The biochemical abnormalities vary between individuals as well as between families, and there is no relationship between the biochemical abnormalities and the blood pressure.

Correction of the biochemical abnormalities by thiazide diuretics is one of the hallmarks of the disease. Thiazides inhibit the reabsorption of sodium chloride (NaCl) by an electroneutral NaCl co-transporter in the collecting tubule. Gordon and Hodsman (1986) reported the immediate return of the biochemical abnormalities, but not of the hypertension, in a patient with Gordon's syndrome following temporary withdrawal of thiazide diuretic after 7 years of treatment. In the patient reported here both the biochemical abnormalities and hypertension returned when thiazide was stopped.

In this patient, bendrofluazide was initially stopped because of hyponatraemia and hypokalaemia as a result of the renal effects of uncontrolled diabetes mellitus. Once the complicating effects of diabetes had resolved bendrofluazide could have been restarted in order to maintain control of the biochemical abnormalities in Gordon's syndrome. Although angiotensin-converting enzyme inhibitors are indicated for diabetic patients, they, as well as

Dr Atef B Michael is Specialist Registrar in the Department of Geriatric Medicine, Queen's Hospital, Burton on Trent, Staffordshire DE13 0RP and **Dr Peter Wallis** is Consultant Physician in the Department of Geriatric Medicine, Birmingham Heartlands Hospital, Birmingham

Correspondence to: Dr AB Michael

potassium-sparing diuretics, are contraindicated in patients with Gordon's syndrome owing to the risk of hyperkalaemia.

Gordon's syndrome is a rare disorder. Since the disease was first described in 1964 more than 90 single cases and families have been reported (Achard et al, 2001). It is not feasible for medical personnel to be familiar with all rare diseases and syndromes, so it is good practice for a warning message to be included in an obvious place in patients' records or notes in GP surgeries and hospitals, giving notice and advice about such rare and potentially serious conditions. Patients with rare disorders should also be informed about their condition and provided with a small card (similar to the familiar steroid card) giving a brief description of the condition. Patients are often best placed to inform doctors about their rare disorders and any contraindicated medications or essential treatments. The internet is also a useful tool for health professionals, enabling rapid access to information about rare conditions. In Gordon's syndrome, the importance of maintaining thiazide treatment and avoiding angiotensin-converting enzyme inhibitors should be emphasized as this case illustrates. HM

The authors would like to thank Dr S Smith, Consultant Nephrologist at Birmingham Heartlands Hospital, for his contribution, and Dr D Milford, Consultant Paediatrician at Birmingham Children's Hospital, for providing the pedigree chart of the family.

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In the Public's View...

Only in my back yard

It's all very well consulting the public, but only if the public are first given the right information. Unfortunately, if the information comes from doctors, it can easily be presented as prejudicing rather than informing.

Bristol has three acute hospitals. It's historical, and it doesn't make sense any more, but the public become attached to 'their hospital' and politicians know it.

The Bristol Royal Infirmary is part of the United Bristol Hospitals NHS Trust, which includes the Bristol Oncology Centre and the Bristol Children's Hospital. The Children's Hospital is only a few years old but was too small when it was built. The site is in the centre of the city, no one can park, and the transport infrastructure in the city is a joke.

Frenchay Hospital looks much as it did at the end of the Second World War. It has long heat-leaking corridors built, I suspect, of corrugated cardboard and asbestos sheeting. On a cold day in winter fog forms inside the corridors. It houses a supraregional neurosurgical centre and a burns unit, and receives major trauma from the nearby motorways. It's impossible to park after 10 o'clock.

Southmead Hospital has a few fine old buildings at its centre, from which, amoeba-like, it has spread. It is a transplant centre and has a large obstetric unit. The old tuberculosis hospital, like many of its kind latterly a hospital for cold orthopaedic surgery, was moved lock stock and staff to rob Southmead of its sports ground and its capacious car park.

With Frenchay, Southmead is one of the two main hospitals of North Bristol NHS Trust. Trust meetings take place in one or other hospital, and you can tell when they're taking place from the people circling the car park searching for a space at whichever hospital is the venue. After the meeting, those same people have to return to the other hospital, and search all over again.

Some of the specialties are split between sites, and divorced from other specialties essential to them. One example is ear, nose and throat surgery (ENT), which is now to be centralized on one site. Except that the children (some say) must still all go to the Children's Hospital. And plans to centralise ENT on one site or another have materialized many times in the past, only for irreconcilable differences, or financial constraints, to force them to be ditched.

And the most important factor is politics. Every time anyone plans anything for the integrated future of Bristol's health services, the local politicians immediately gain populist support by howling that 'their' hospital is to be disadvantaged. Here's a Conservative newsletter (but the others are no different) promising to carry out a survey of local views but while the public may want a hospital at the end of every street, super-specialization and too few doctors to staff on-call rotas mean that it just isn't possible.

Whatever is decided for Bristol after this latest round of consultation and feasibility studies, the earliest that any new hospital can open its doors is 2012. My interest has changed from looking forward to working in a streamlined modern hospital system to needing treatment in one. HM

Dr Neville W Goodman is Consultant Anaesthetist at Southmead Hospital, Bristol