SELF ASSESSMENT QUESTIONS

Obstetric difficulties due to Graves' disease

A Bhattacharyya, J D Wright, P A Vice

Answers on p 669.

A 14 year old girl was referred to the paediatricians with symptoms of hyperthyroidism. She had a smooth diffuse goitre with dysthyroid eye disease (proptosis, lid lag, and lid retraction). Hyperthyroidism was confirmed biochemically (protein bound iodine 18.8, normal 5–8 µg). She was treated with carbimazole, 30 mg/day. Poor compliance resulted in inpatient care for treatment with carbimazole followed by subtotal thyroidectomy, two years later. Two years later she presented with a self limited episode of hyperthyroidism. Aged 22 years she was referred with eight weeks amenorrhoea when pregnancy was confirmed and terminated. She was floridly hyperthyroid (free thyroxine 36.6, normal 10–23 pmol/l, free triiodothyronine 14, normal 3–9 pmol/l; thyroid stimulating hormone (TSH) undetectable, normal 0.5–5 mU/l). She was treated with carbimazole for nine months with apparently good clinical response.

At 24 years, she was admitted in premature labour at 36 weeks, resulting in a stillborn infant (weight 1700 g, goitre of 4.4 g, and diffuse hyperplasia on histology). No obvious maternal thyroid problem had been noted during the pregnancy. Six months later an unplanned pregnancy was terminated at 10 weeks of gestation when she was proved to be hyperthyroid (free thyroxine 30 pmol/l). At 26 years, she had a first trimester spontaneous abortion. A further pregnancy at the age of 30 years without obvious thyroid disease was complicated by premature labour, resulting in an urgent caesarean section for fetal distress and a stillborn baby. This baby weighed 1870 g and a goitre was noted (thyroid weighing 6 g, diffuse hyperplasia on histology). It was when she was readmitted with wound infection that she was noted to be hyperthyroid and was referred to the Division of Endocrinology.

She was clinically and biochemically hyperthyroid (free thyroxine 23 pmol/l and free triiodothyronine 11.3 pmol/l) with dysthyroid eye disease. Her TSH receptor antibody (TSHRAb) was 95 (normal 0–10 U/l), thyroglobulin antibody (TGA) was 1: 81 920, while thyroid microsomal antibody (TMA) was

1: 1 638 400 (normal being less than 1: 400 for both TGA and TMA). The patient wished to try for a future pregnancy. After discussion, it was agreed that she would receive an ablative dose of radioactive iodine (with contraceptive care after the dose), to render her hypothyroid and maintain her on thyroxine. The plan was successful and she became hypothyroid within three months of receiving radioactive iodine. She was euthyroid on 100 µg thyroxine daily. Fifteen months after receiving radioiodine she conceived and was seen in the Department of Obstetrics by JDW. She was closely monitored throughout the pregnancy. TSH and thyroxine confirmed she remained euthyroid throughout the pregnancy. TSHRAb became undetectable in late pregnancy (table 1). Fetal monitoring confirmed an euthyroid state (no tachycardia) with normal growth and no goitre on the scan. Amniocentesis, done to assess the maturity of the fetal lungs (this is not the practice now) at 36 weeks, was complicated by antepartum haemorrhage. Emergency caesarean section resulted in a live baby (birth weight 3400 g, no goitre, normal thyroid function, and negative TSHRAb). She conceived again at 35 years, resulting in a further successful outcome (birth weight 3700 g). She was managed in the same way as the previous successful one (the TSHRAb remained in the normal range throughout the pregnancy), the baby was euthyroid, and there was no goitre. She was sterilised after the second childbirth.

Questions

- (1) Would more aggressive monitoring of the maternal thyroid status and fetal condition in the unsuccessful pregnancies have led to a better outcome?
- (2) Is it likely that the difference in the outcome of those unsuccessful pregnancies was related to the maternal uncontrolled thyroid state or to the circulating antibody levels?
- (3) What are the three crucial factors for the successful outcome in this case?

Table 1 Showing the thyroid parameters, antibody status, and treatment during the first successful pregnancy

	Normal values	Preconception	First trimester	Second trimester	Third trimester	Two months after delivery
Free thyroxine (pmol/l)	10–23	12.9	17.5	15	21.9	15
Free triiodothyronine (pmol/l)	2–5.5	4.1	4.7	3.1	3.9	4.2
TSH (mU/l)	0.5–5	1.6	4.1	7.4	0.9	1.5
TSHRAb (U/l)	0–10	40	21	17	5	5
Daily dose of thyroxine (µg)		100	100	200	200	100

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A rare presentation of a common disease

H Patel, K S Hindle, G Tsavellas, A Huang

Answers on p 670.

A previously well 77 year old man presented with acute left iliac fossa pain. He was pyrexial and tachycardic with localised abdominal guarding but there was no palpable mass or generalised peritonitis. Respiratory examination revealed tachypnoea with normal bilateral air entry and no tracheal deviation. Blood investigations showed a leucocytosis with raised serum C reactive protein and erythrocyte sedimentation rate. Serum amylase was normal and he was hypoxic with reduced oxygen saturation in the blood. His abdominal and

Figure 2 Erect thoracic radiograph.

Figure 1 Abdominal radiograph.

Figure 2 Abdominal computed tomogram.

erect thoracic radiographs are shown (figs 1 and 2). He also underwent abdominal computed tomography (fig 3).

Questions

- (1) What are the radiographic findings?
- (2) What is the most likely diagnosis?
- (3) What are the treatment options?

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Adrenal mass in a diabetic with hypergastrinaemia

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Answers on p 671.

A 50 year old man originally presented in 1980 at the age of 31 with diarrhoea, weight loss, and abdominal pain. He was initially treated with steroids for presumed inflammatory bowel disease. About a month later he presented with severe abdominal pain, and a laparotomy revealed multiple jejunal perforations and a possible mass in the head of the pancreas. He made a good recovery during which time a

nasogastric tube was in place for two weeks. A markedly raised serum gastrin level confirmed the diagnosis of Zollinger-Ellison syndrome. Other gut hormone levels were normal. A parathyroid adenoma was suspected on account of raised calcium and parathyroid hormone levels.

He underwent a parathyroidectomy, and since then his serum calcium has remained normal.

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Biopsy of the nodule in the head of the pancreas confirmed a pancreatic islet cell tumour. He subsequently underwent total gastrectomy, distal oesophagectomy for a peptic oesophageal stricture, and pancreaticoduodenectomy in 1980.

His gastrin levels fell after surgery, but rose again sharply within the next two years.

He remained well until 1995, when he presented with weight loss, polyuria, and polydipsia.

Diabetes mellitus was diagnosed and he was started on insulin therapy. At this time a computed tomography scan of his abdomen revealed a 4 × 2 cm right adrenal mass and a 1.5 cm nodule in the tail of the pancreas. An octreotide labelled scan was normal, and the adrenal mass was thought to be an incidental mass. Subsequent computed tomography indicated

that the adrenal mass was growing in size, and was removed surgically in 1999. Histology was diagnostic of primary adrenal carcinoma.

Questions

- (1) What other conditions besides Zohlinger-Ellison syndrome could cause a raised gastrin level?
- (2) Why does hypercalcaemia sometime present with abdominal pain?
- (3) How would you proceed to investigate whether the adrenal mass is functioning or just an incidental finding?
- (4) Why did this man develop diabetes mellitus?
- (5) Why is it important to screen his family?
- (6) Why did he develop a distal oesophageal stricture?

Adult intussusception—an elusive diagnosis

W J Sotheran, M H Wise

Answers on p 672.

An 81 year old woman presented, as an emergency, with a two day history of abdominal pain. The pain was intermittent and confined to the right upper quadrant. She had no other symptoms and normally enjoyed good health. Examination revealed tenderness in the right hypochondrium. Investigations revealed a mildly raised serum alkaline phosphatase. All other haematological and biochemical parameters were within normal limits. A provisional diagnosis of biliary colic was made.

Four days later, the patient experienced diarrhoea and vomiting. Her abdominal pain had increased and there was generalised abdominal tenderness. She developed a pyrexia of 38°C. An ultrasound scan demonstrated a distended gall bladder with stones and a dilated common bile duct (12.6 mm). Antibiotic treatment was started. The patient made little progress with continued diarrhoea and vomiting. Stool cultures and sigmoidoscopy were normal. Parenteral nutrition was started.

Over the next two days, the patient developed a palpable mass in the epigastrium. Repeat ultrasound showed a normal biliary tree, but a mass consistent with a necrotic pancreas with pseudocyst was imaged. Computed tomography of the abdomen and pelvis was requested. Computed tomograms are shown in figures 1 and 2.

Questions

- (1) The images show a characteristic mass lesion anteriorly. What diagnosis is suggested by these findings?

Figure 1 Contrast enhanced computed tomogram at the L2 level showing an abnormal soft tissue mass in the right side of the abdomen.

Figure 2 Another view from the same examination shows the same abnormal mass central and anteriorly.

- (2) How may this condition present in adults?
- (3) How should such cases be managed?

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Acute respiratory failure in a middle aged woman

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Answers on p 673.

A 61 year old woman presented with a one week history of headache, drowsiness, and shortness of breath. Initial examination revealed bilateral ptosis, which the patient stated had been present for three to four years before this admission. She also reported that her father had died suddenly, shortly after developing "drooping eyelids". There was restriction in the range of eye movements in all directions. The remaining cranial nerves were intact.

In the limbs there was normal tone but mildly reduced power in all muscle groups, worse proximally. There was generalised areflexia. Plantar responses were flexor and there was no sensory deficit. Coordination was normal. There were no signs of meningism. On chest examination there was reduced air entry bilaterally and bronchial breathing in both lower zones. Cardiovascular and abdominal examinations were unremarkable.

There was a past medical history of pernicious anaemia, osteoporosis, partial thyroidectomy, and cholecystectomy. Drugs on admission were disodium etidronate and hormone replacement therapy.

Laboratory investigations included an arterial pH of 7.34, oxygen pressure 11.78 kPa, carbon dioxide pressure 7.1 kPa, and a base excess +5.1. Urea and electrolytes, creatinine, calcium, creatinine kinase, and lactate were all within normal limits. Full blood picture was unremarkable and cerebrospinal fluid examination was also normal. A chest radiography showed bilateral lower lobe collapse and consolidation. The electrocardiogram is shown in fig 1.

The patient's condition deteriorated with increasing respiratory distress. Her respiratory rate rose to 32 breaths/min, repeat arterial blood gases indicated pH 7.30 kPa, oxygen pressure 9.9 kPa, and carbon dioxide pressure 8.9 kPa. The patient was transferred to intensive care for ventilatory support.

Questions

- (1) What is the differential diagnosis for this clinical presentation?
- (2) What does the electrocardiogram (fig 1) show and what is the significance of this?
- (3) What other clinical findings would be useful to elicit?
- (4) What further investigations would you like to perform?

Figure 1 Electrocardiogram.

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Answers on p 675.

Cor pulmonale: variation on a theme

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A woman aged 89 was admitted with a three month history of effort dyspnoea. She had had a previous admission, at the age of 86, for left lower lobe pneumonia, with coincidental atrial fibrillation.

On examination she was centrally cyanosed, normotensive, and had atrial fibrillation, pedal oedema, and raised jugular venous pressure to the angle of the jaw even when sitting up. A soft systolic murmur was audible at the left sternal edge.

Questions

- (1) What investigations would help to identify the underlying cause of cardiac failure in this patient?
- (2) Could there be a link between the atrial fibrillation and evolution of the changes in the QRS axis?

Rectal bleeding in a patient with portal hypertension

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Answers on p 677.

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A 77 year old woman presented for the first time in 1997 with severe haematemesis and melena. After initial resuscitation she underwent an emergency upper gastrointestinal endoscopy which showed bleeding oesophageal varices. She was treated with injection sclerotherapy.

She had a history of ischaemic heart disease but was stable on medication. She was not known to have any other medical problems.

Clinically she was pale but not jaundiced. There was mild ascites and prominent abdominal veins. The splenic tip was just palpable below the left costal margin. Her liver function tests were normal. There was no coagulation abnormality. Ultrasound scan of the abdomen showed normal hepatic architecture. The spleen was enlarged and there was evidence of ascites. Portal blood flow studies suggested portal hypertension secondary to thrombus in the portal vein. After discharge from the hospital she was kept under endoscopic surveillance. The variceal recurrences needed repeated ligation.

Three years later she had to be admitted again, this time for the investigation of bleeding per rectum. It was intermittent, self limiting, and small in amount. Her bowel habits were normal. Clinical examination revealed pallor, mild ascites, and prominent abdominal veins. Her spleen was palpable 2 cm below the costal

Figure 1 Finding on flexible sigmoidoscopy.

margin. Figure 1 shows the finding on flexible sigmoidoscopy. Her barium enema was normal.

Questions

- (1) What was the most probable cause of rectal bleeding in this woman?
- (2) What is the prevalence of this condition in portal hypertension?
- (3) What is the management?

A state of confusion

N Sofat, C S Higgins

Answers on p 677.

A 66 year old right handed man was admitted via the accident and emergency department with confusion and weakness. His wife gave most of the history, saying he had not been able to express himself clearly for the last day and looked weak. He had no history of ischaemic heart disease, hypertension, hypercholesterolaemia, diabetes mellitus, or previous strokes. He smoked 10 cigarettes a day and drank alcohol only socially. He had been diagnosed with rheumatoid arthritis 20 years before. His drug history included diclofenac. On examination, the admitting doctor found that he had an expressive dysphasia and a right hemiparesis. He had a blood pressure of 140/70 mm Hg, pulse 80 beats/min and regular, both first and second heart sounds present with no added sounds and his chest was clear. His abdomen was soft and non-tender with no organomegaly. He had bilateral metacarpophalangeal joint swelling in his hands with ulnar deviation. On further neurological assessment he had an upper motor neurone right facial nerve palsy and the rest of his cranial nerves were normal. In his limbs, in addition to the right hemiparesis, he also had an ulnar nerve palsy in his left hand.

The patient was diagnosed as having had an acute stroke. Computed tomography the same day showed areas of hypodensity consistent with fresh infarctions in the temporal lobes bilaterally and also in the left frontoparietal lobe. He was started on 300 mg aspirin daily. His plasma lipids and glucose were found to be normal. Doppler examination of his carotid arteries and cardiac echocardiogram were also normal. He was transferred to the neurorehabilitation unit. Five days after his admission, he was noted to have a lesion in his left eye as illustrated in fig 1. He was also seen to have a lesion over his left lateral malleolus, shown in fig 2, which was very painful.

Figure 1 Illustration of patient's eye as seen a few days after admission (reproduced with patient's permission).

Figure 2 Photograph of the patient's lower leg (reproduced with patient's permission).

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Questions

- (1) What is illustrated in fig 1?
- (2) What is the skin lesion illustrated in fig 2?
- (3) What is the unifying diagnosis?
- (4) What treatment would you now offer the patient?
- (5) What is his prognosis?

Answers on p 678.

Dysphagia in a patient with palmoplantar keratoderma

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A 72 year old man was referred by his general practitioner with a four month history of dysphagia. Physical examination was

unremarkable with the exception of his hands and feet which showed hyperkeratosis on the palms (fig 1) and soles of his feet. Gastroscopy showed severe extensive oesophagitis (grade III) confirmed with multiple oesophageal biopsies. He was started on a proton pump inhibitor (omeprazole) and rescoped three months later. He still had oesophagitis at this time, although macroscopically it had improved; repeat biopsies again showed oesophagitis. A further gastroscopy three months later showed complete resolution of his oesophagitis confirmed by repeated oesophageal biopsies. He remains well and asymptomatic on omeprazole and continues to be followed up.

Figure 1 Hyperkeratosis on the patient's palms.

Questions

- (1) What is the diagnosis?
- (2) How is it inherited?
- (3) What is the major complication?

Decreased sexual function in a young man

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A 36 year old professional man presented with an 18 month history of difficulty in achieving and maintaining an erection. He also described "lack of sex drive", malaise, and non-specific ill health over several months. He was sometimes able to masturbate successfully but commented that his ejaculate was of small volume. Physical examination showed him to be anxious but he was otherwise normal except for the fact that the testes were smaller than expected (about 12 ml in volume) and soft.

Questions

- (1) What is the commonest cause of erectile dysfunction in a man of this age?
- (2) What features of the clinical presentation in this patient would be in favour of an organic cause for his problem.
- (3) What baseline investigations would you undertake?

Answers

Q1: What is the commonest cause of erectile dysfunction in a man of this age?

Erectile problems in young men are, on average, more likely to result from psychological than physical causes,¹ though a combination of the two may be present. The older the

patient at presentation, the more likely is the disorder to have a mainly physical cause.

Q2: What features of the clinical presentation in this patient would be in favour of an organic cause for his problem

The reduction in libido, small ejaculate volume, and small, soft testes are all suggestive of hypogonadism.

Q3: What baseline investigations would you undertake?

In this case there is clear evidence of the need to measure serum testosterone, prolactin, follicle stimulating hormone (FSH), and luteinising hormone. In many clinics a baseline blood glucose and renal and liver function tests would also be requested.

The patient's initial endocrine results are shown in table 1. Plain skull radiography and

Table 1 Patient's endocrine results

		Reference range
Testosterone (nmol/l)	1.6	>10.0
Free androgen index	4	>14
FSH (U/l)	1.4	2.0–12.0
Luteinising hormone (U/l)	2.8	1.0–12.0
Prolactin (mIU/l)	1063	<555

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Figure 1 Plain skull radiograph.

Figure 2 MRI scan.

magnetic resonance imaging (MRI) were requested (figs 1 and 2).

The low serum testosterone and free androgen index confirm hypogonadism.

Questions

- (4) Is this patient's hypogonadism primary or secondary? Why?
- (5) What is the abnormality seen on the skull radiography (fig 1)?
- (6) What diagnosis is suggested by the MRI scan (fig 2)?
- (7) At what age does this condition typically present?
- (8) What treatment should this patient have for his primary disorder?
- (9) What treatment should he have for his hypogonadism? Why?

Answers

Q4: Is this patient's hypogonadism primary or secondary? Why?

This patient has secondary hypogonadism: his serum testosterone is very low but there is no compensatory increase in the secretion of FSH and luteinising hormone—in a man with primary testicular failure and a serum testosterone of this level one would expect the serum FSH and luteinising hormone to be greater than 30.

Learning points

- Erectile dysfunction in younger men is commonly caused by mainly psychological factors but organic factors should nevertheless be sought.
- The presence of loss of libido or of small, often soft, testes favours a diagnosis of hypogonadism.
- If the serum testosterone is low and the levels of FSH and luteinising hormone are low or normal then the patient has secondary hypogonadism due to pituitary or hypothalamic disease.
- Craniopharyngiomas may present in adulthood.

Q5: What is the abnormality seen on the skull radiography?

There is suprasellar calcification.

Q6: What diagnosis is suggested by the MRI (fig 2) scan?

The MRI scan shows a solid soft tissue mass in the suprasellar cistern. It does not appear to arise from the pituitary and is most likely to be a craniopharyngioma.

Q7: At what age does this condition typically present?

Craniopharyngioma is most commonly detected between the ages of 6 and 14 but may come to light in adults, even up to an advanced age.^{2,3}

Q8: What treatment should this patient have for his primary disorder?

The patient will require a full pituitary assessment, including a combined pituitary function test. The lesion itself is compressing the optic chiasm and will require surgery followed by radiotherapy.

Q9: What treatment should he have for his hypogonadism? Why?

The patient should be offered a choice of forms of testosterone replacement, both for the sake of his sexual function and in order to preserve his muscle mass and prevent osteoporosis. Available treatments include intramuscular testosterone esters, oral testosterone undecanoate, transdermal testosterone, and testosterone implants.

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