testosterone. The increase in their activities may lead to a low testosterone concentration and a high dihydrotestosterone concentration. These changes may explain the reduced ratio of testosterone to dihydrotestosterone found in patients with pancreatic cancer.

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Isolated central respiratory failure due to syringomyelia and Arnold-Chiari malformation

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We report on two patients without pre-existing neurological abnormality who presented to hospital with acute central respiratory failure. Both showed syringomyelia and an Arnold-Chiari malformation on magnetic resonance imaging, and both improved after decompression of the foramen magnum. Isolated respiratory failure has not to our knowledge been reported in association with syringomyelia.

Case reports

Case 1—A healthy 26 year old woman became breathless over two weeks. She became lethargic, tended to fall asleep at work, and finally was found unconscious in bed. On admission she was cyanosed, hypothermic, and unconscious. Analysis of blood gases while she was breathing air showed a pH of 6.82, arterial carbon dioxide tension of 22.9 kPa, and arterial oxygen tension of 5.3 kPa. After intubation and ventilation she became fully conscious. Neurological examination showed brisk reflexes and inadequate spontaneous ventilation; her arterial carbon dioxide tension rose to 8.0 kPa over six hours. A computed tomogram was normal, but magnetic resonance imaging showed an Arnold-Chiari malformation with syringomyelia extending from the medulla to the thoracic spinal cord (figure). Bilateral paralysis of the

diaphragm was confirmed by radiography, and she communicated that she was afraid to fall asleep without the ventilator in case she stopped breathing. Capnography and spirometry showed that arterial carbon dioxide tension increased and ventilation decreased when she fell asleep (Ondine's curse). Decompression of the posterior fossa, duroplasty, and aspiration of the syrinx were performed. Within 10 days she was able to breathe spontaneously when awake but required ventilation to sleep. Though she needed long term nocturnal ventilation through a close fitting nasal mask, she continued to improve and returned to work.

Case 2—A healthy 58 year old woman became confused over two days. She stumbled and struck her head against a table but did not lose consciousness. She was referred for neurosurgical assessment because of confusion. No skull fracture or neurological abnormality was present, but analysis of blood gases when she was breathing air showed a pH of 7.33, arterial carbon dioxide tension of 8.8 kPa, and arterial oxygen tension of 7.6 kPa. A computed tomogram showed a shallow posterior fossa, and magnetic resonance imaging showed a syrinx extending from the medulla to the conus. Spirometry showed a reduction in vital capacity. After decompression of the posterior fossa and duroplasty her arterial carbon dioxide tension fell to 5.3 kPa and arterial oxygen tension to 8.0-8.6 kPa on air. She became fully lucid and more active than before admission.

Comment

Central respiratory failure has been described in neonates in association with Arnold-Chiari malformation, hydrocephalus, and spina bifida, and decompression of the foramen magnum has been recommended when lower cranial nerve or respiratory dysfunction occurs.¹ Central respiratory failure has occurred in association with Arnold-Chiari malformation and with syringomyelia and lower cranial nerve palsies,² and in one case respiratory failure was attributed to insensitivity of peripheral chemoreceptors resulting from lesions of the ninth cranial nerves.³

Both our patients had large syrinxes, yet neither exhibited the classic symptoms or signs of syringomyelia before presenting with acute respiratory failure. In both cases radiographs of the diaphragm confirmed phrenic nerve dysfunction, which improved after operation. Phrenic motor neurons are located more medially in the anterior horn than other motor neurons and may be vulnerable to compression by an expanding syrinx.

Central respiratory failure in these patients may have been due to compression of the medulla by the Arnold-Chiari malformation or to the syringomyelia. The effect on the phrenic nerve suggested that the syringomyelia was important, although Arnold-Chiari malformation alone has caused respiratory failure.⁴

Both patients were diagnosed by magnetic resonance imaging, and in one case the computed tomogram was normal. Magnetic resonance imaging of the cranio-vertebral junction should be considered in patients

Sagittal magnetic resonance images before and after operation (case 1), showing a large syrinx that collapsed after surgery and descent of tonsils to level of C1

who present with respiratory failure of uncertain origin, even when no other neurological abnormality is present.

We thank Dr M Braithwaite, Brompton Chest Hospital, London, for help with case 1.

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Detection of lupus anticoagulant in patients attending an anticoagulation clinic

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The presence of the lupus anticoagulant is now known to indicate a tendency towards thromboses and recurrent abortions.¹ In patients with a history of thrombosis identifying the presence of lupus anticoagulant is clinically important as long term anticoagulation treatment should perhaps be considered. We determined the prevalence of lupus anticoagulant in selected patients attending an anticoagulation clinic.

Patients, methods, and results

About 1300 patients in total attend the anticoagulation clinic at this hospital. The following patients were selected to be screened for lupus anticoagulant: patients of any age with a history of recurrent thrombosis or an episode of thrombosis at a site other than veins of the legs, such as the axillary vein; patients aged less than 40 with a history of venous or arterial thrombosis in the absence of any known predisposing factors, such as the use of contraceptive pills, prolonged immobilisation, malignancy, obesity, or cardiac arrhythmias; and women with a history of both thrombosis and recurrent abortions or unexplained intrauterine death. The

presence of lupus anticoagulant was detected by measuring the effect of patients' plasma on the kaolin clotting time of normal plasma.² The patients were also screened for anticardiolipin antibodies.

We identified 29 patients who fulfilled the above criteria, comprising 12 men and 17 women with a median age of 36 (range 22-60). Nine of these patients (four men, five women) were found to be positive for lupus anticoagulant (figure) on two separate occasions. The 20 other patients were negative for it. Of the nine patients with lupus anticoagulant, three had a history of arterial thrombosis, of whom one had had two myocardial infarctions. Four had a history of recurrent venous thrombosis, and two had a history of both venous thrombosis and recurrent abortions. Seven were tested for anticardiolipin antibodies and five were found to have raised titres. One patient had coexisting thrombocytopenia, but none had any clinical history or laboratory features suggestive of systemic lupus erythematosus.

Comment

Our results show that the selection criteria that we used will identify a subpopulation of patients attending an anticoagulation clinic who have a surprisingly high prevalence of lupus anticoagulant (30%). This could be of considerable clinical value as the presence of lupus anticoagulant is increasingly appreciated as constituting a risk factor for thrombosis. Possibly such patients should be given prophylactic long term anticoagulation treatment.³ In the case of pregnant patients such information would form a rational basis for advice on treatment. We emphasise, however, that our study was targeted at a highly selected group of patients, and the overall prevalence of lupus anticoagulant in unselected patients with thrombosis is as low as 2%.⁴

In conclusion, we recommend that screening certain selected patients attending anticoagulation clinics will identify several for whom long term anticoagulation treatment might be an advantage.

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Positive result of testing for lupus anticoagulant (○-----○), showing convexity on left, and limits of negative results obtained in six unselected patients treated with warfarin (●—●)

Correction

Vasoactive and atherogenic effects of cigarette smoking: a study of monozygotic twins discordant for smoking

An editorial error occurred in this paper by Dr R Lassila and others (15 October, p 955). In the third sentence of the third paragraph of the discussion prostaglandin $F_{1\alpha}$ receptor was substituted for α_1 adrenoceptor. The sentence should have read: "In humans vasoconstriction mediated by α_1 adrenoceptor releases urinary prostaglandins in response to infusion with norepinephrine,¹⁴ and in an animal model stimulation of the sympathetic nerves leads to a significant rapid increase in output of prostacyclin from the mesenteric arterial bed."¹⁵