Convulsion and coma after intraocular desmopressin in cystic fibrosis

Dr E J Simmonds, M J Mahoney, and J M Littlewood (Regional Cystic Fibrosis Unit, St James's Hospital, Leeds LS9 7TF) write: Desmopressin has been advocated as a treatment for nocturnal enuresis.1 We report on a child with cystic fibrosis who developed water intoxication while being treated with desmopressin.

She was 10 years old and had cystic fibrosis with nasal polyps. She had had polyp surgery at the age of 1 year, and had been taking fenbufen (130 mg three times a day). Pancreatic and vitamin supplements were being given at a concentration of 2190 IU/1 (230-460), alkaline phosphatase 1233 IU/1 (100-280), and albumin 30 g/l. A Paul-Bunnell test, autoantibody screen, and toxoplasma antibody titres were all negative. Enzymatic sedimentation rate, chest radiograph, radiograph of the left knee, and calcium and phosphate concentrations were normal.

Four days later she had a deep jaundice due to severe hepatitis. Repeat blood cultures were negative. Her haemoglobin started to drop and was 5 g/l 10 days after stopping fenbufen and was treated with 2-4 g/l, prothrombin time 2-4 s, activated partial thromboplastin time 41 s (control 21-32), fibrinogen normal. Aspartate aminotransferase activity was 26 IU/l, alanine aminotransferase 39, asialojiggluzaminase 2190 IU/1 (230-460), alkaline phosphatase 1233 IU/1 (100-280), and albumin 30 g/l. A Paul-Bunnell test, autoantibody screen, and toxoplasma antibody titres were all negative. Enzymatic sedimentation rate, chest radiograph, radiograph of the left knee, and calcium and phosphate concentrations were normal.

This patient with cystic fibrosis had a severe adverse response to intraocular desmopressin used in doses recommended by the manufacturer; several authors have reported that desmopressin by this route is safe for both short1 and long term2 treatment of nocturnal enuresis. There are no reports of water intoxication, although the basic defect in a few patients treated with intranasal desmopressin for diabetes insipidus,3 it is not clear why our patient responded as she did. As we have no other cases of severe physiological adverse effects on the liver.

Four days later she was deeply jaundiced with persistent fever. Repeat blood cultures were negative. Her haemoglobin started to drop and was 5 g/l 10 days after stopping fenbufen and was treated with fenbufen 900 mg/day for a painful left elbow. She took it for 10 days, and four days after stopping the drug she developed a rapid rise in liver enzymes and became unwell with thirst, pain, fever, and rigors. There was no history of antihistamine, drug allergy, a change in temperature, or an alteration in her well (temperature 40°C) with generalised lymphadenopathy and an extensive erythematous exfoliative rash affecting the whole body and face. The left knee was swollen and tender. Investigations showed: haemoglobin 133 g/l, white cell count 42 × 10⁹/l, eosinophils 10 × 10⁹/l, neutrophils 14 × 10⁹/l, platelets 139 × 10⁹/l; alkaline phosphatase 319 IU/1 (20-150), gamma glutamyl transferase 2190 IU/1 (230-460), alkaline phosphatase 1233 IU/1 (100-280), and albumin 30 g/l. A Paul-Bunnell test, autoantibody screen, and toxoplasma antibody titres were all negative. Enzymatic sedimentation rate, chest radiograph, radiograph of the left knee, and calcium and phosphate concentrations were normal.

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