gestive of the 3 conditions reported as being a cause of progressive subglottic narrowing in adults.² Most reported cases of perichondritis with subglottic involvement have had associated inflammation of the nasal cartilages. Sarcoidosis is a rare disorder in children and usually there is generalised lymphadenopathy or changes on the chest radiograph. A diagnosis of Wegener's granulomatosis was entertained in Case 4 but there were no other features of the disorder although the boy did have granulomatous panniculitis. The relationship of this and of Hashimoto's disease in Case 3 to the subglottic lesion is unclear. There is a single case report⁸ of an adult with progressive non-traumatic subglottic narrowing but this did not respond to steroids.

It is important that paediatricians be aware of this disorder. These children all had findings typical of laryngeal obstruction although for months they were thought to have asthma, and were dangerously obstructed by the time they were finally diagnosed. The disorder appears to be very sensitive to corticosteroids and these should be used in patients suspected of having a similar condition before resorting to major surgery.

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Spontaneous hypothermia in a young boy

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SUMMARY A 13-year-old boy who maintains his body temperature between 33 and 35°C was investigated. Tests of peripheral and central thermoregulatory function did not show a specific abnormality, except for some generalised hypothalamic dysfunction of unknown aetiology. He responded appropriately when his temperature was altered from its usual low level. His temperature regulatory mechanisms appeared to be set at a temperature lower than is normal.

Hypothermia is defined as a rectal or body-core temperature below 35°C. Accidental hypothermia is common—especially in the elderly¹—but spontaneous hypothermia is rare—particularly in

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young adults—if hypothyroidism is excluded. Several cases have been reported during the last 50 years,^{2–5} but studies of underlying central and peripheral thermoregulatory function are few, especially in children. We report results of investigations on a hypothermic 13-year-old boy.

Case history

The boy was born in India, a term normal delivery, birthweight 3.2 kg. He developed jaundice in the first 3 days of life which persisted for about 4 weeks. No blood tests were done or treatment given. He breast fed well and there were no other illnesses. He sat at age 8 months and walked at 19 months. He can communicate and read in his native language

and perform daily chores but is thought by his family to be slow. He speaks only a few words of English. The parents and 6 siblings are healthy. He came to the UK in September 1980 and presented 2 months later with a history of withdrawal and slowness, particularly since his arrival here.

On examination his oral temperature was 33.8°C, pulse rate 72/min, and regular; heart sounds normal, blood pressure 110/60 mmHg, chest clear, abdomen normal. Head circumference was 52.1 cm, cranial nerve examination and fundoscopy were normal. It was possible to elicit a weak triceps reflex but other reflexes could not be elicited. Plantar reflexes were flexor and sensation to temperature (cold and warm), touch, pressure, pain, and position appeared normal. There was no loss of muscle power and autonomic responses appeared normal. Response to Valsalva's manoeuvre was normal and there was no hypotension on tilting. Weight and height were appropriate for age and parental sizes. He was thought initially to be myxoedematous but thyroid function tests gave normal results. He was therefore, further investigated and observed.

Investigations. The following investigations were normal: blood count, urea, and electrolytes, erythrocyte sedimentation rate, liver function tests, blood sugar, creatine phosphokinase, serum cholesterol, triglyceride, glucagon, autoantibody screen, immunoglobulins, serum thyroxine (156 nmol/l), tri-iodothyronine (2.2 pmol/l); skull and chest x-ray films, electrocardiogram, Mantoux, Wassermann reaction, computerised tomography scan of head. Electroencephalogram showed non-specific slow wave changes; electromyogram showed normal conduction velocity but a small sural sensory action potential. Examination of the cerebrospinal fluid showed no cells and a protein concentration of 0.54 g/l; no evidence of virus infection was found. Serology was negative for influenza, mumps, measles, adenovirus, cyto-

megalovirus, herpes, psittacosis, Q fever, and mycoplasma. There was no specific rubella IgM antibody and toxoplasma dye test was normal.

A combined insulin tolerance, thyroid-releasing hormone, luteinising hormone-releasing hormone test was carried out to test hypothalamo-pituitary function. Hypoglycaemia produced a poor and late response to adrenocorticotrophic hormone, which was reflected in a poor cortisol rise. The thyroid-stimulating hormone response to thyroxine-releasing hormone was inadequate and the prolactin response was rather flat. The gonadotrophin responses and diurnal variation of the hormones were normal. A synacthen test showed a normal adrenal response to injected adrenocorticotrophic hormone, serum cortisol rising in 30 minutes from 212 to 800 nmol/l.

Diurnal urine excretion and a water deprivation test were normal. Muscle biopsy was histologically normal, and the mitochondria were normal in structure, numbers, and enzyme function.

In the ambient temperature of the hospital ward (21–22°C) his oral and rectal temperature remained between 33 and 35°C.

In response to cooling (Table 1) he was able to shiver and his basal metabolic rate rose. Basal metabolic rate was assessed by the open circuit method of indirect calorimetry, using a Douglas bag. During the same period the temperature of a control patient fell by 0.4°C; she shivered more and appeared to be more uncomfortable than the patient. In response to warming (Table 2) he started feeling uncomfortable before the control patient and this discomfort increased as the temperature rose. At 37.3°C he became agitated and uncooperative whereas the control experienced only mild discomfort although her temperature rose to 37.4°C. The patient's basal metabolic rate increased only slightly and he did not show the usual correlation between respiration rate, pulse rate, and body temperature. No formal pyrogen test was done but his temperature rose to 35.3°C during a common cold.

Table 1 *Effect of cooling*

Time (hours)	Temperature (°C)			Heart rate (per minute)	Respiratory rate (per minute)	Comments	Basal metabolic rate (KJ/m ² per hour)
	Room	Skin	Oral				
1110	23	29	33.6	58	11	Resting	253
1140	10	26.5	33.6	60	12	Complains of feeling cold. Control 37°C	
1200	10	23	33.2	70	12	Shivering, goose pimples	236
1230	10	21.8	32	58	12	Occasional ectopic beats	
1250	10	21.0	31.8	48	12	No more ectopic beats. Control 36.8°	324
1310	10	21.0	30.6	44	12	Shivering less	
1325	10	20.5	31	42	15	Alert, notices nurses looking through window.	426
						Feels cold and hungry. Control 36.6°	
1340	10	20	30	42	12		

Table 2 Effect of warming

Time (hours)	Temperature (°C)			Heart rate (per minute)	Respiratory rate (per minute)	Comments	Basal metabolic rate (KJ/m ² per hour)
	Room	Skin	Oral				
0930	23	29	32.8	53	13		352
1000	26	30.6	33	55	14		
1030	31	32.9	33.2	52	13	Feels uncomfortable	
1100	34	34	33.4	63	10		
1130	35.5	34.3	34	66	13		
1200	40	35.7	34.9	70	12	Feels uncomfortable	
1230	41	36	35.2	78	11		
1300	47	36.4	36.6	87	12		
1325	43	36.7	37	102	13	Sweating, very uncomfortable	
1345	42	37	37.3	108	12		383

Discussion

The cause of this boy's hypothermia remains unknown. His temperature-regulating mechanism appears to be set at a lower level than is normal but he responds appropriately although in an exaggerated manner when his temperature is altered from its usual low level. In normal people a drop in central temperature below 36°C will cause shivering, even in the absence of stimuli from cold skin,⁶ but in this patient shivering occurred at a much lower (33.2°C) temperature.

Hypothalamic and central nervous system dysfunction is a known cause of hypothermia.^{2 4 5} In this patient computerised tomography scans of the head failed to show a lesion affecting the hypothalamus. The insulin hypoglycaemia test is the best test for assessing the functional adequacy of the hypothalamic-pituitary-adrenal axis,^{7 8} and in this patient the results of the insulin tolerance test suggested hypothalamic dysfunction. There was no evidence of syphilis, tuberculosis, sarcoidosis, or a slow virus infection—such as kuru or scrapie. The most likely cause of this boy's hypothermia is a very small lesion of the anterior hypothalamus which is not yet detectable on computerised tomography scanning.

The patient and his family have been advised to ensure that he has adequate insulation especially of the extremities to prevent heat loss, particularly in

cold weather. He will be followed up closely as an outpatient.

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