

alone was administered by intermittent positive-pressure respiration. This made no difference whatsoever to the inexorable progress of the condition.

Details of time, temperature, and acid-base state are presented in Tables I and II.

TABLE I.—Temperature Changes (Degrees Centigrade)

	Minutes after Induction						
	0	15	30	45	60	90	120
Pig 1	*—	—	—	—	44.0	43.4	—
Pig 2	*—	—	—	—	38.0	41.2	42.4
Pig 3	38.0	38.0	40.0	42.0	43.5	44.5	44.5
Pig 4	—	39.0	41.0	42.0	43.0	—	—
Pig 5	—	38.0	42.5	43.0	—	—	—
Pig 6	—	39.0	42.0	>45.5	45.5	45.0	45.0
Time of death				Pig 5 (45 min.)	Pig 4 (60 min.)	Pig 1 (105 min.)	Pigs 3 and 6 (120 min.) Pig 2 (135 min.)

\* Early monitoring of temperature was undertaken only after our first experiences of the condition.

TABLE II.—Acid-base Studies\* (Femoral Arterial Blood—Astrup Technique)

		Minutes after Induction			
		30	60	90	120
Pig 1	pH	—	6.85	—	—
	P <sub>CO<sub>2</sub></sub>	—	130	—	—
	B.E.	—	-16.8	—	—
Pig 2	pH	—	7.37	7.12	6.85
	P <sub>CO<sub>2</sub></sub>	—	40	80	144
	B.E.	—	-1.5	-5.9	-19.0
Pig 3	pH	—	—	7.08	7.1
	P <sub>CO<sub>2</sub></sub>	—	—	55	67
	B.E.	—	—	-8.4	-7.7
Pig 5. Mixed venous blood	pH	6.65	—	—	—
	P <sub>CO<sub>2</sub></sub>	90	—	—	—
	B.E.	> -22.0	—	—	—
Pig 6. Mixed venous blood	pH	6.44	6.73	6.79	6.99
	P <sub>CO<sub>2</sub></sub>	> 150	> 150	71	136
	B.E.	> -22	-4.0	+7.1	-1.5

\* (a) Studies were begun after femoral arterial cannulation. Pig 4 succumbed before this event. Because of this an early mixed venous specimen was taken from Pig 5. (b) The values shown for Pig 3 reflect the acid-base state after the administration of 200 mEq of sodium bicarbonate. Between the 30-minute and 120-minute periods in Pig 6 a total of 375 mEq of sodium bicarbonate was given.

## DISCUSSION

Malignant hyperpyrexia during anaesthesia has been described in both humans and pigs. A new aspect of this problem that emerges from our report is that it can occur in the absence of the use of suxamethonium, the drug on which other workers have focused attention. Examination of the recently published reports of malignant hyperpyrexia during anaesthesia shows that, with one exception, the common

factors to all these cases and our own are the anaesthetic agents—a fluorinated hydrocarbon and nitrous oxide. The mechanism by which so immense an amount of heat is so rapidly produced poses a fascinating physiological problem. Does some disturbance of oxidative phosphorylation play a part (Snodgrass and Piras, 1966; Wilson *et al.*, 1966)? Viguera and Conn (1967) failed to produce this picture of malignant hyperpyrexia in chickens, both by the production of muscle spasm and by dinitrophenol-induced uncoupling of oxidative phosphorylation. Even more fundamental than the problem of the mechanism of heat production is the question of the precise identity of the stimulus that initiates this veritable metabolic storm. Most important to the solution of this problem is the salvaging of one of the pigs that react in this way so that we may see if the event is experimentally reproducible.

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## Medical Memoranda

### Congenital absence of the Lumbar Spine

*Brit. med. J.*, 1968, **3**, 595-596

Complete absence of the lumbar spine is a rare congenital anomaly. When it occurs it is associated with a flaccid paraplegia and disturbance or absence of sphincter control which result in incontinence of faeces and urine. The presence of such disabilities and the absence of part of the spine have in the past been regarded as a hopeless situation. As a result such patients have lived in institutions for the physically handicapped. No active measures have been taken to enable them to walk or be useful members of the community. We have had several such cases and we intend to report the findings and results of treatment in these at a later date. At this stage we

simply describe one of our earlier cases to draw attention to the fact that such patients can be enabled to walk and become more happy and useful members of society.

## CASE REPORT

The patient was born on 15 January 1945 and was soon found to have paraplegia, together with rectal and urinary incontinence. Because of his gross disability he was brought up in a hospital for crippled children. Radiographs showed absence of the lumbar spine (Fig. 1).

When seen here at the age of 17 years he could only sit like Buddha and move around by "walking" on his hands. He was able to do this because of the disproportionately small abdomen and very large chest and the strength and length of his arms (Fig. 2).

After consultation at this hospital bilateral through-knee amputation was performed elsewhere in March 1963. The wounds healed by primary intention and he was returned to Roehampton for limb fitting. Clinically the problem was how to fit leg prostheses to a

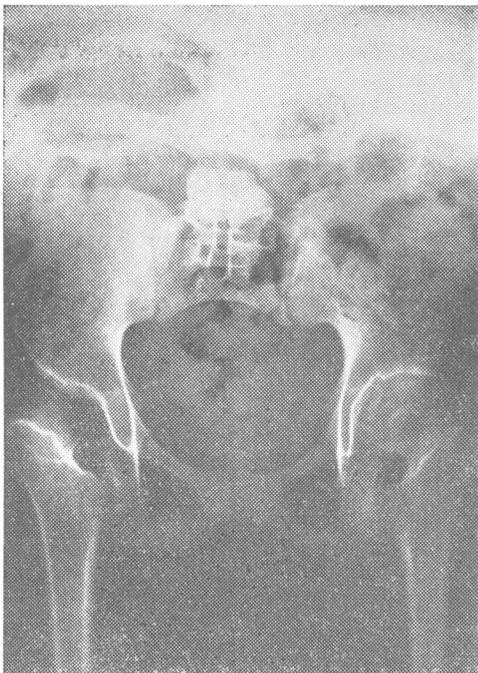


FIG. 1.—Radiograph showing absence of lumbar spine.

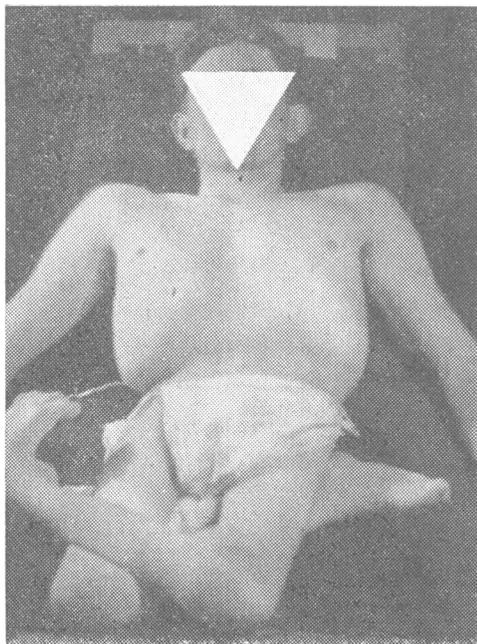


FIG. 2.—Photograph of patient at 17 years of age.

man who had two good but anaesthetic and flail through-knee stumps, whose hip joints had a very limited range of movement, and who had no lumbar spine to give rigidity to the lower part of the trunk. Because of the latter, when he sat he flexed at the level of the spinal gap and tended to "fold up" on himself.

The prosthetic rehabilitation programme was divided into two phases: (1) fitting a pair of short, temporary, non-articulated pylons on which the patient could learn to walk and test the proposed method of fixing; and (2) fitting a pair of permanent articulated limbs, so providing both mobility and cosmetic rehabilitation.

*Phase 1.*—Pylons were made with wooden rocker ends and light-weight rigid metal struts on which were mounted metal sockets fitted with soft end-bearing pads. A blocked leather corset was made from a plaster cast taken of the chest from the costal margin

to the axillae. This chest corset was mounted on the pylons by means of side struts jointed at the level of the gap between the twelfth dorsal and the sacral block of bone. The prostheses were arranged so that about half of the weight was taken on the chest corset and the other half shared between the gluteal seats and the end-bearing pads of the sockets. The patient learnt to balance on these limbs without much difficulty and was able to walk, using two sticks. After a few weeks the pylons were lengthened to increase his height.

*Phase 2.*—About three months later work was started on the fitting of permanent articulated prostheses. The bag for his urinary appliance was accommodated within the right shin. The increased weight and height of the permanent limbs caused some difficulty at first, but after further instruction in the walking-school these problems were overcome and he walked quite well with both knees locked and using two sticks (Fig. 3).



FIG. 3.—Patient with articulated prosthesis fitted.

At follow-up six years later he was wearing his artificial limbs about three times a week, sometimes in the evenings only. He walked well and could manage up to 100 yards (90 metres) using his sticks. He worked sitting, in the packing department of a factory. He drove a motor-propelled invalid carriage to and from work. He also visited friends many miles from home. He usually travelled in his vehicle without his artificial limbs, because he could then more easily swing himself from the driver's seat to a collapsible wheelchair (which is attached to his invalid car). He was living in a hostel for invalids, for which he paid board and lodgings. He was saving money and expected to get married in the next few years and buy himself a house or flat. He considered himself independent. His bowels were well controlled, and lately he had even learnt to control his urine. Mentally, he was much brighter and more mature.

*Comment.*—It is to be hoped that any other such patients, whatever their age, are referred to centres where help and advice can be given. Our results in other cases suggest that it would be helpful to begin treating children as soon as they are about 2 years of age.

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