Conclusions

Partial rebreathing is justified in so far as it promotes lung expansion and oxygenation, and retains the warmth and moisture of the upper respiratory contents while permitting adequate ventilation below.

Its extent is modified in the continuous-flow machine by adjustment of the gas flow, and in the intermittent-flow machine by adjustment of the bag capacity.

The patient's effective alveolar ventilation is controlled, as always, by the alveolar CO_2 concentration.

For continuous-flow machines there is a "critical flow" in the neighbourhood of 5 litres per minute for each individual, which if diminished will lead to CO_2 accumulation, but which if exceeded allows equilibrium to be attained.

Continuous flow is dangerous with a flow below the critical level; intermittent flow is dangerous with an unlimited bag capacity.

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LAURENCE-MOON-BIEDL SYNDROME, WITH TETANY

BY

FRANCIS D'ABREU, M.Ch., F.R.C.S.

Major, R.A.M.C.

AND

DAVID FERRIMAN, D.M., M.R.C.P.

The following interesting case, considered to be an example of the Laurence-Moon-Biedl syndrome without polydactyly or mental deficiency, seems to merit publication.

Case History

A young woman aged 25 was admitted to Westminster Hospital in October, 1938. Her immediate complaint was of pain in the left loin that had been present for a week. The features which had actually led to her admission were blindness—present from birth—and attacks of tetany. These attacks dated from the age of 10, and had been occasional until nine months before. From that time they became more frequent, and just before her admission were occurring about twice a week. During the last two years she had also complained of pain in her back, for which she had been given a spinal jacket at another hospital. A report from this hospital stated that radiographs of the bones seemed normal. There was no history of vomiting. She had at one time been rather fat; with the onset of tetany she had gone to the opposite extreme, but recently she had returned more or less to normal. Her menstruation had been irregular and infrequent from the onset at 11, but the intervals had steadily increased from the minimum of three months, her last period having taken place four years ago. A further sixteen sibs of this patient had been born, of whom only five are now alive. Four of these sixteen were blind at birth, and only one is living. No abnormal fatness, tetany, or mental deficiency had affected any sibs; a sister with unaffected vision, however, had had six toes on either foot—she died early in life. The mother suffered from diabetes, but otherwise she and the father were healthy; they were unrelated.

A number of typical attacks of tetany with carpo-pedal and facial spasm were observed during the patient's stay in hospital. Chvostek's and Trousseau's signs were positive and

could be elicited at any time with ease. Her veins were abnormally irritable: none were visible and no blood could be obtained from any of them, not even after cutting down and exposing them, as they went into intense spasm on the prick of the needle. Eventually blood had to be taken by needle from the femoral artery. The nursing staff had not observed any hyperpnoea. Mr. E. F. King examined the patient's eyes, and important findings are as follows: No light perception; pupils inactive to light; posterior cortical complicated cataracts; both retinæ show gross primary degeneration with secondary narrowing of the retinal vessels and optic atrophy. "The picture is somewhat like retinitis pigmentosa, though the size and distribution of the pigmentation are not typical, and of course the history of blindness from birth is atypical." The patient was plump but hardly obese; polydactyly was not present. Her breast development and hair distribution were healthy. She appeared quite intelligent. She was tender in both loins and the urine contained albumin.

Special investigations gave results as follows: Serum calcium 9.2 mg., 10.9 mg., and 9.3 mg. per 100 c.cm.; plasma phosphorus 3.8 mg. and 4.2 mg. per 100 c.cm.; serum phosphatase 11 units; alkali reserve 61 c.cm. carbon dioxide %; blood urea 19 mg. per 100 c.cm. Radiographs of lumbar and dorsal spine disclosed nothing abnormal. The urine contained *B. coli*. An intravenous pyelography failed to reveal abnormality. The basal metabolic rate was +14%. The glucose-tolerance curve was normal, and the Wassermann and Kahn reactions were negative. A full blood count was normal.

The patient was put on three-quarters of an ounce of calcium lactate and 50,000 international units of vitamin D daily. Dr. G. A. Tuck, her medical practitioner, reports that she is greatly improved and that she has had only one attack of tetany in the last three months. It is interesting that her veins are now readily visible.

Discussion

Both the cause of the tetany and the diagnosis of the case are obscure. The investigations appear to rule out any of the accepted causes of tetany. An explanation was somewhat tentatively advanced by Dr. Raymond Greene. It runs as follows:

"Obesity has been present at one time; this and the amenorrhoea might be of pituitary or hypothalamic origin. A parathyrotrophic hormone is to be found in the pituitary. The increasing intervals between the periods and the variation in the obesity suggest an alteration in the level of pituitary secretion over a period of years. If the secretion of the parathyrotrophic hormone at some earlier time was high but has now sunk to normal levels (as indicated by present normal blood calcium and phosphorus figures) then it is possible that the tetany of this case is a consequence of the body's failure to adapt itself, as yet, to the relatively low calcium level."

The result of treatment with calcium and vitamin D supports an explanation along some such lines. No other hypothesis has been suggested.

We believe this case to be an example of the Laurence-Moon-Biedl syndrome. Admittedly polydactyly and mental deficiency are absent, but this has been so in other accepted cases. The absence of obesity is unusual, though Cockayne and co-workers (1935, 1939) have found records of this in a few cases; further, obesity was at one time present in this case. The menstrual disturbance supports the diagnosis. The obesity and amenorrhoea of the syndrome have usually been taken to indicate a pituitary or hypothalamic lesion. A lesion of such a nature in this case is further suggested by the tetany. Cockayne, Krestin, and Sorsby (1935) say, in connexion with atypical retinitis pigmentosa in this disease, that "typical retinitis pigmentosa would appear to be the exception." The affection of four other sibs, the parents being unaffected, is suggestive of a recessive mode of inheritance, and this is consistent with what is known of the syndrome (Cockayne and co-workers, 1935, 1939). The appearance of poly-

dactly in another otherwise unaffected sib is significant and also rather typical (Cockayne and co-workers, 1935, 1939).

No other diagnosis has been advanced.

This case is described by the courtesy of Mr. G. T. Mullally, under whose care it was.

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Clinical Memoranda

A Suspected Case of Chronic Meningococcal Septicaemia

Details of the following case are submitted because it is felt that the diagnosis of chronic meningococcal septicaemia is easy if the condition is borne in mind, and because, with the present high incidence of cerebrospinal fever, there must be many cases which pass unrecognized.

Chronic meningococcal septicaemia was originally reported by Saloman (1902), and it has since been seen in increasing numbers on the Continent and in America. The only case reported in England since Stewart-Wallace (1936) recorded 3 from the London Hospital was that by Dimson (1938), which was treated with sulphapyridine. Stott and Copeman (1940) analysed seventeen cases out of a total of twenty-seven which they had seen among members of the B.E.F. in France. Stott (private communication) saw a considerable number more after the paper had gone to press, but had not seen any in this country. As pointed out by Appelbaum (1939), the condition occurs among both sexes and in all age groups. It is common before and after an epidemic of cerebrospinal fever, and is often ushered in by an acute tonsillitis or other upper respiratory infection.

CASE HISTORY

The patient was a previously healthy soldier aged 24 who had never been out of the country. He was admitted to hospital on September 5, complaining of sore throat and fever. His temperature that night was 103° and his pulse 108; his throat was red and injected. A throat swab yielded an abundant growth of haemolytic streptococci. By the following day his temperature was normal, and he made an uninterrupted recovery; so much so that he was due for discharge from hospital on September 12, when he had his first bout of fever.

On the evening of the 12th he retired to bed with a temperature of 103°, sweated, felt cold, and said that his head ached. On the following morning his temperature was normal, but a crop of spots was noticed on his legs, arms, and back. These spots were hard, raised, macular, and tender, and recurred in crops; they resembled slightly those seen in erythema nodosum. His temperature rose again on the evening of the 13th, but by the following morning was again normal: the cutaneous eruption had faded, leaving slight brownish pigmentation. On the 15th, 18th, 21st, 25th, and 26th his evening temperature rose to 102–104°, his pulse rate rarely reached 100 and never exceeded 120. A fresh crop of spots appeared with each bout of pyrexia and faded in about thirty-six hours. At no time did the patient feel ill, he was not clinically anaemic, and the spleen was not palpable.

A radiograph of the sinuses on September 11 revealed slight clouding of both antra, suggesting catarrh. A radiograph of the chest on September 20 showed the lungs to be clear. Blood cultures on the 21st, 25th, and 27th were sterile. Examination of a film of blood on the 25th revealed no malarial parasites, and the blood Wassermann reaction was negative.

A full course of sulphapyridine was begun on the 27th, and thereafter the temperature remained subnormal. The patient was discharged from hospital on October 8.

This case is interesting from the point of view of the diagnosis. Chronic meningococcal septicaemia is probably

commoner than is supposed, and may become increasingly so because of the high incidence of cerebrospinal fever which has been maintained throughout the year. Its clinical recognition should be simple, according to Stott and Copeman (1940): pyrexia, associated with a relatively slow pulse, a cutaneous eruption which may be pleomorphic but is often similar to that of erythema nodosum, and the absence of malaise, wasting, and anaemia are the main points which they stress. Blood cultures in the above case were negative, and in Stott and Copeman's series there were only six positives. Cerebrospinal fluid is always normal in the uncomplicated cases. Response to sulphapyridine is immediate.

The focus of infection is as yet unknown, but it is possible that sinus infection plays a part. Rolleston (1919) quoted a case with antral infection, but pus obtained by puncture did not yield meningococci. In my case the antra were cloudy, but, as this was attributed to catarrh, puncture was not carried out.

I am indebted to Dr. L. C. Martin, physician to the hospital, for permission to publish the details of this case, which was under his care, and for help in preparation. Also to Colonel A. W. Stott, R.A.M.C., consulting physician to the Forces, for help in the final diagnosis of the case.

A. GRAHAM PORTER, B.A., M.B., B.Ch.,
House-physician, Emergency Hospital, Cambridge.

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The Pectoralis Major for Intramuscular Injections

The buttock, though universally recommended, has many drawbacks as a recipient of intramuscular injections. The fat-screen is variable and often massive; the marks and guides are indefinite; there are large veins, arteries, and nerves within reach (I have more than once known the sciatic nerve injected); if tenderness develops from the injection, walking, standing, sitting, and even lying down are painful. The region is apt to have unaesthetic associations, there is often hesitation in giving sufficient exposure, and falls of drapery may be embarrassing. These considerations many years ago impelled me to seek another site, and the edge of the pectoralis major was chosen; it has been used almost exclusively since, and no drawbacks have been found.

The doctor stands at the side of the patient, preferably the left. Men patients remove their shirt from that shoulder; women may usually retain their underclothes if these have fairly large armholes. The patient raises the arm sideways to a right angle; resting the hand on the doctor's right shoulder often gives confidence, and the slight pull on the arm brings the edge of the pectoralis major into prominence. After cleaning the skin with antiseptic the doctor's left thumb is placed on the axillary side of the muscle and his forefinger in front of it; the edge of the muscle is thus grasped, and a little pressure will make it tense and facilitate the puncture. The needle is inserted parallel with the shoulders and with the anterior surface of the muscle, between the thumb and finger, which serve as guides.

It is easy to direct the needle into the muscle with precision, and its penetration can be felt. There is little fat in the way, there are no large vessels or nerves in the neighbourhood, and it is not very sensitive. Ten c.cm. is accommodated with ease, 20 c.cm. is well tolerated, and absorption is rapid. The same site can be used repeatedly, though it is easy to choose different parts of the muscle. Bismuth and other opaque injections can easily be observed with x rays, and their absorption so watched. Pain or tenderness is rare; even with irritating materials they appear to be less frequent than in the buttock, possibly because extra-muscular injection is less often achieved. If tenderness should develop the left arm can usually be rested or even placed in a sling.

Birmingham.

CRANSTON WALKER, M.D., B.Sc.