Some aspects of the management of diabetic coma in small babies have been discussed. These include: the value of 1/6 molar sodium lactate solution; some suggestions on insulin dosage; frequency of hypoglycaemia due to prolonged insulin action; hypernatraemia; and hypokalaemia.

The importance of parental intelligence and co-operation in subsequent management is stressed.

I thank Dr. J. F. Landreth, senior paediatrician to the Christchurch Hospital, for permission to report these cases and for his help and encouragement; and Mr. J. T. Murray, biochemist, for his enthusiasm for electrolyte estimations at all hours and for first drawing my attention to some of the problems of acidosis and of hypernatraemia in diabetic coma.

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AETIOLOGY AND TREATMENT OF CONGENITAL DEAF-MUTISM

BY

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During an investigation into the problem of congenital abnormalities of the head it was noted that occasionally a child showing obvious maldevelopment of the first visceral arch was also deaf-mute, although the external ear and auditory meatus were normal. Attention was thereby drawn to the possibility of a close connexion between deaf-mutism and abnormal development of the first arch.

The cases described below show little or no obvious evidence of first-arch defect, and the above premise suggested the clinical experiment of determining whether their deaf-mutism was associated with middle-ear (first arch) defect.

Case 1

A boy aged 11 was suspected at the age of 2 years of having middle-ear deafness, "possibly from infection in the first few months of life." When he was first examined it was reported that he did hear—cochleo-palpebral reflex was present but weak. The surgeon had the impression when using tuning-forks that it was a conduction deafness, but was not at all certain. Although six months later (aged 2½) the child showed signs, with pure-tone bone-conduction audiometry, of a severe upper-tone loss in both ears, a tentative diagnosis of air-conduction deafness associated with mental deficiency was made.

At 3½ years co-operation was said to be non-existent during clinical examination; a muscular tic reported a year previously was more pronounced. No final diagnosis was reached, and special schooling was recommended. No other member of the family suffered from deafness or congenital

abnormality, and x-ray examination of the skull was normal. Fig. 1 shows the result of audiometry examination (May, 1957); deafness is shown to be profound, but the marked discrepancy between air-conduction and bone-conduction hearing indicates that at the lower frequencies the deafness is essentially of middle-ear origin, although the upper-tone loss suggests inner-ear deafness.

Case 2

A girl aged 9½ was found most difficult to examine at the age of 2, when the cochleo-palpebral reflex was elicited on very slight stimulus. At 5 years of age the only finding of note was a single islet of hearing at 1,024 cycles revealed by pure-tone audiometry. She was a pleasant, but very shy, timid, little girl, and of her relatives only an aunt of her mother's was reported to have been born deaf;

no further information was available. The patient was consigned to a school for the deaf without further attempts to arrive at the cause or nature of the deafness. At school she was given and wore an air-conduction hearing-aid as a routine, but it was not regarded as being of much help to her. The skull was normal on x-ray examination. Audiometry (Fig. 2) revealed a deafness strikingly similar to that of Case 1.

Cases 3, 4, and 5

Three children aged 12, 9, and 9, respectively (the latter two being twins) were all members of one family suffering from a congenital deafness, which must be hereditary. The mother was born deaf, but the trait could not be traced further back on her side of the family. The father was one of five deaf-mute sons in a family of nine; one of his affected brothers died in a mental hospital, where another was still an inmate; the others and the father of the three patients were normal apart from the deafness. X-ray

FIG. 1.—Case 1. O = Right. X = Left.

FIG. 2.—Case 2.

FIG. 3.—Case 3.

examination of the skull was normal in all three cases. Audiometry examination (Figs. 3, 4, and 5) was very similar in each case to those of Cases 1 and 2. There had been no previous otological or audiometry examination of these

FIG. 4.—Case 4.

FIG. 5.—Case 5.

children, and all have been wearing air-conduction hearing-aids at the school for the deaf.

Careful attention was given to the possibility of the child confusing hearing with vibration during bone conduction testing, and in each case we were satisfied that the patient was able to differentiate them clearly.

Discussion

No one has ever suggested that congenital deaf-mutism may be of middle-ear origin; in fact, the condition is generally regarded as an affliction of the internal ear. Wilson (1955) deals very briefly with congenital abnormalities of the internal

ear, merely saying, "This subject is inextricably mixed up with deaf-mutism, to the chapter on which the reader is referred"; in that chapter he mentions only once the involvement of the middle ear in cases of congenital deaf-mutism—namely, as an incidental finding when the cause is syphilis, and that, he says, is infrequent.

Recently there has been an awareness that few children are completely deaf. The majority of those recognized as deaf-mutes are classified as "profoundly deaf," and schools, unfortunately with different principles of teaching, have been established for these children. Speech-training at first meant finger-spelling, but very soon attempts to teach them to speak were begun. Lip-reading was encouraged, but efforts have more recently been directed towards making the children hear as much as possible in their training. All children attending these schools use hearing-aids, either individual or group models of the air-conducting type.

The results of this investigation are surprising and unexpected. Testing by air conduction showed profound deafness, while by bone conduction there was remarkably good hearing at the lower frequencies, with no response whatsoever to the higher. There is undoubtedly a marked internal-ear or perceptive deafness, but the picture is dominated by the degree of air-conduction deafness, a feature so far unsuspected by clinician or teacher.

The full significance of this discovery is, first, that congenital deaf-mutism is not due entirely to an internal-ear or perceptive deafness, and, second, that if these children are to be expected to hear as much as possible then bone-conduction hearing-aids should be used. One great objection to this type of aid is the size and weight of the batteries supplied for the instrument, but there appear to be no technical difficulties in producing more suitable batteries. The third point also concerns the treatment. From bone-conduction audiometry it is obvious that there is an appreciable cochlear defect—almost normal hearing at the lower tones

"tailing off" to give no response at the upper tones; the loss is neither irregular nor patchy. The difference between air- and bone-conduction readings points to gross middle-ear involvement and shows that it is the more serious, although, clinically, the middle ears of such patients had been regarded as normal.

The problem being one of deciding which is the primary defect, three reasons are offered for regarding the middle ear as being initially at fault. (1) Two congenitally deaf-mute children, unfortunately too young to co-operate in the examination, showed obvious deformities of the first visceral arch, and this arch contributes to the formation of the middle-ear structures. (2) The middle-ear deafness was greater in all those examined, and it is unlikely that a less degree of perceptive deafness could cause a major degree of conduction deafness. (3) While the cochlea and middle ear are closely associated functionally, the structural association between the cochlea and vestibular apparatus is even greater; yet in children with congenital deaf-mutism it is unknown to find lack of balance due to coincident involvement of the vestibular apparatus (except in cases of syphilitic infection). If the defect were primarily labyrinthine and secondarily middle ear, then surely signs of involvement of the vestibular part would be found in at least some of the cases, especially those who are absolutely deaf.

No morphological evidence of a defect of the middle ear in these cases is available; x-ray examination with various projections has been of no assistance in identifying the presence or absence of the ossicles, far less their normality, but the difficulties of radiodiagnosis in this region are well recognized (Hodgson, 1951).

Whether the congenital abnormality of the middle ear is minimal or gross, its effect is clearly that of interrupting the transmission of vibrations to the internal ear. A developmental defect such as this, arising in the early weeks of intra-uterine life, must have an effect on the associated parts of the hearing mechanism, and it may be best expressed as a loss of stimulation to further development of the inner ear. Whether this loss takes effect during the remainder of intra-uterine life, thereby leading to structural maldevelopment, or whether the effect occurs in the equally obscure and unknown period of the development of hearing—namely, in the first few months of life—is a matter for conjecture, but it is much more likely to be after birth, when the initial abnormal changes will be functional rather than structural. Thus the pathological processes are in the nature of a disuse atrophy of the cochlea. Accordingly, not only must the training of the deaf-mute comprise the use of a bone-conducting aid, but it must also start as early as possible, at a stage when it may still be possible to retrieve leeway lost in the first few months of life.

Even if all cases of deaf-mutism do not arise from primary defects of the middle ear, the disadvantage of fitting each child with an elaborate hearing-aid and awaiting the result of audiometry examination at the age of 7 or 8 is far outweighed by the advantages which are sure to accrue from the earliest stimulation of what hearing exists. By this means it is likely that little, if any, perceptive hearing loss (except perhaps in the highest tones) will be present by the time the child is old enough to be examined properly; and in the meantime it is likely that speech will have advanced to nearly normal standards.

The interrelationship of a wide field of congenital abnormalities of the head and neck will be described in a subsequent publication, but the findings in regard to congenital deaf-mutism were considered important enough to warrant this preliminary report and a discussion on its aetiology.

Summary

The results of audiometric examination in five deaf-mute children are presented. In each case the degree of hearing by bone conduction is much greater than that by air conduction, an indication that the primary defect is

the result of congenital maldevelopment of the middle ear and not of the inner ear, as is generally supposed. In order to stimulate and improve the residual hearing in these children treatment should therefore begin at the earliest possible age with the use of a bone-conducting hearing-aid.

I am greatly indebted to Professor R. D. Lockhart for his encouragement and advice during this investigation, to Mr. I. S. D. Thomson for allowing me access to his notes on Cases 1 and 2, and to Miss J. C. Macrae for carrying out all the audiometry examinations.

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HYDATID COMPLEMENT-FIXATION TEST: INFLUENCE OF INTERMEDIATE HOST PROTEIN ON SPECIFICITY OF SHEEP HYDATID FLUID ANTIGEN

BY

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Kellaway and Fairley (1932) considered the hydatid complement-fixation test to be specific, and we (Bensted and Atkinson, 1953), reporting on the examination with standardized reagents of a small series of cases, fully supported this opinion. Since that time a further series of over 300 cases of suspected hydatid disease have been examined serologically by the same technique, and out of 70 cases, subsequently proved to be hydatid disease, significant complement-fixing antibody was demonstrated in the serum of 61 of the patients concerned. There were 240 patients in whom further investigation showed that their condition was due to some disease other than hydatid. Sera from all but one of these patients gave negative results in the hydatid complement-fixation test. It is this false-positive result—the first in our experience—that initiated the investigations recorded in this communication. The explanation proved to be simple, and once the facts were known the results of the investigations were in full accord with what was expected. Nevertheless some interesting observations were made which seemed worthy of record as they had not, so far as is known, previously been published.

During radiological examination the patient in question had been found to have a suspicious shadow in the lung, and blood serum had been forwarded for the C.F.T. to exclude hydatid disease. The Casoni test had been reported as negative, but the C.F.T. with the standardized sheep hydatid fluid antigen was positive to a titre of 1 in 16. Further clinical investigations, however, revealed that the patient had a carcinoma of the lung. The disease ended fatally in a short time and no evidence of a hydatid cyst was found.

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It was at first thought that the complement fixation might have been due to the development of some non-specific reacting substance associated with the rapidly growing neoplasm, but our records showed that 12 similar cases of carcinoma of the lung had been examined serologically for the presence of hydatid complement-fixing antibody and in all the test had been completely negative. Further inquiries, however, revealed that the patient had recently returned to this country from the Middle East, where, a few weeks before sailing, he had been bitten by a rabid dog and had been given a "full course" of a sheep-brain anti-rabies vaccine.

Some time after this incident, and additional to the series referred to above, a similar case came to light. In this case, however, the anti-rabies vaccine had been given some years previously. Two Casoni tests gave atypical delayed reactions, and the C.F.T. performed after a short interval showed partial fixation at a titre of 1 in 2 only. The case subsequently proved to be carcinoma of the lung.

Without the history of the previous case these results would doubtlessly have been disregarded, but now the possibility of sheep-protein sensitization from the sheep-brain anti-rabies vaccine and the boosting effect of the intradermal injections of the sheep hydatid fluid (for the Casoni tests) seemed to warrant further investigation.

Although there was little doubt that individuals recently immunized with sheep-brain anti-rabies vaccine would have some sheep-protein antibodies in their sera it would be necessary to check this. There was considerable delay before we were able to obtain material for such tests, and in the meantime we decided to try to obtain the required evidence from animal experiments.

Experimental

Rabbits

Rabbits are not suitable for immunization experiments with sheep-tissue vaccines. Friedemann (1917) had shown that rabbits possess a natural haemolysin for sheep's red blood cells, and by the use of suitable antigens it was found that other sheep antibodies, such as complement-fixing antibodies for sheep protein, could be demonstrated in the sera of a high proportion of normal rabbits. The presence of this natural complement-fixing antibody in the serum of normal rabbits suggested that further examination of this point might yield information of interest.

Hiles (1926) had noted that the sera of normal rabbits gave positive complement fixation with sheep hydatid fluid and assumed that the reaction was due to some anti-hydatid substance in the sera. We confirmed Hiles's observations, but when we tested a representative number of sera against ovine and bovine hydatid fluids† and a suspension of normal sheep's brain in parallel, we found that practically all the sera showed some degree of fixation of complement with the sheep-brain antigen, but that only those sera with a relatively high anti-sheep titre gave fixation with an ovine hydatid fluid. Reactions with the bovine hydatid fluid were only occasionally seen. When complement fixation occurred with the sheep hydatid fluid the titre was significantly lower than that against the sheep-brain antigen; the occasional reactions with the bovine fluid were incomplete. Rabbit sera did not react with pure hydatid antigen, and absorption of a serum, giving fixation with both sheep-brain and sheep hydatid antigens, with massive doses of washed scolices from a fresh hydatid cyst failed to reduce the complement-fixation titre to either antigen. The titre of a serum containing true hydatid antibody was reduced to nearly zero in a parallel absorption test.

†Diluted in accordance with assay requirements.