A deaf-mute with Huntington's cholera

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Huntington's cholera was said to have originated from an infidel who mocked by mime Christ's agony on the Cross (Bickford and Ellison, 1953). Most descendants of this man have been spared other afflictions: the disease is horrible enough. Rosanoff (1923), Hughes (1925), Spillane and Phillips (1937), and Lyon (1962) have all commented on the rarity of any associated nervous or mental disease. Thus Rosanoff found that other conditions occurred 'relatively so infrequently as to be readily accounted for as coincidences essentially without relation to the chorea itself'.

Congenital deafness always carries the risk that the sufferer will never acquire speech and will consequently be forced to sign his wants. Such people, even today, may justifiably be referred to as deaf-mute. That a person relying on gesture for all communication should also be afflicted with the dread chorea must appear as a singularly cruel quirk of fate.

This patient, we believe, is only the second to be reported with this association of disorders. In the earlier case report (Bonduelle, Gruner, and Bougues, 1953) no objective reference was made to their patient's difficulty in communication.

CASE REPORT

R.P., a carpenter aged 50, was admitted to this hospital as an emergency in May 1965. He was deaf and dumb. His history was obtained from his wife, who was also a deaf-mute, and interpreted by his 11-year-old daughter.

For six months he had been getting increasingly short of breath, and had had several episodes of abdominal pain and vomiting. He was constipated, but had also been incontinent of faeces. His appetite was poor.

On examination, he showed evidence of dementia and chorea, with poor attention span, frequent grimacing, and continual choreiform movements. The movements involved all four limbs (the arms being more affected than the legs), and the head and neck. He made occasional grunts but was otherwise silent.

The cranial nerves, with the exception of the eighth nerve, were normal. There was no response to loud noises in either ear. Both drums were normal.

Apart from the involuntary movements and postures, motor testing in the limbs was normal, reflexes brisk and symmetrical and plantars flexor. Sensory testing was limited but no abnormality was found.

On admission, his temperature was 99.8°F., pulse 132 per minute, regular, and blood pressure 180/100 mm. Hg.

The heart sounds were normal and there were no signs of heart failure. He was dyspnoic: respiratory rate 32/min. There was no cyanosis. The percussion note was diminished at both bases with absent breath sounds. Bowels sounds were audible at both bases.

The tongue was dry. The abdomen was markedly distended and generally tender. No organs were palpable. The abdomen was tympanitic to percussion and the bowel sounds were tinkling. Rectal examination revealed a large quantity of faecal material.

A diagnosis of Huntington's chorea was made, with dementia leading to chronic constipation, steadily increasing faecal impaction, overflow incontinence, dilatation of the large and small bowel, elevated diaphragm and hence dyspnoea.

Investigations showed haemoglobin 98% (14.5g.%), white blood count 9,300/c.mm., E.S.R. 48 mm. at one hour. Urine microscopy was normal. The plasma urea level was 34 mg.%. Plasma electrolytes: total CO₂ capacity 29.5 mEq./l., chloride 93 mEq./l., sodium 133 mEq./l., potassium 4.4 mEq./l.

Total serum proteins were 60 g.% (albumin 2.8 g.%, globulin 3.2 g.%).

Serum flocculation tests showed colloidal red very weakly positive, cephalin cholesterol weakly positive, and thymol turbidity 2 units. Plasma glutamic oxaloacetic transaminase was 16 Karmen units/min./ml., and plasma alkaline phosphatase 20-3 King-Armstrong units.

A chest radiograph showed that both hemidiaphragms were elevated, but no focal lung lesion was seen. Abdominal radiographs showed gross faecal retention in the descending colon and rectum, and considerable distension of the rest of the large bowel and also of the small bowel.

An electroencephalogram, following the administration of 100 mg. each of butobarbitone and quinalbarbitone and 2 g. of chloral, showed that there was a considerable amount of low-voltage fast activity with bursts of mostly frontal, but occasionally occipitally distributed, activity at between 12 and 14 c/s. These were symmetrical. Some were undoubtedly sleep spindles, but others represented a basic activity just above the alpha range. Only a little generalized intermediate slow activity of very low voltage was seen. Considerable agitation masked the responses
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FIG. 1. Pedigree of the patient.

them is in a mental institution. His mother died, aged 50, in a mental institution after three years of choreiform movements; and his maternal grandmother was also affected.

Little is known of the earlier members of the family, and despite the help of Dr. J. D. Spillane and Dr. C. E. C. Wells in Glamorgan, we have been unable to trace these early sufferers.

DISCUSSION

The cause of this patient’s deafness has not been determined. In two recent surveys of hereditary deafness (Fisch, 1964; Fraser, 1964) no mention is made of any relationship between Huntington’s chorea and deafness, nor had either of these authors encountered such a case in the course of their experience of deafness (personal communications).

Worster-Drought and Allen in 1929 described a deaf and dumb child of a choreic father but did not follow her for sufficiently long to exclude chorea. Minski and Guttmann in 1938 in a survey of Huntington’s chorea note in family 27 of their series ‘several cases of deafness not coinciding with the chorea’. In the absence of further elaboration it may be assumed that these were of adult, acquired deafness. In 1953 Bonduelle et al., in reporting two siblings with Huntington’s chorea and spasmodic paraplegia, mention that they had difficulty communicating with the second patient as she had been deaf and dumb since early infancy. The cause of her deafness had not been determined and no other member of the family suffered from deafness.

Several methods of communication may be used by profoundly deaf people. That of speech and speech-reading (lip-reading) is preferred often to the exclusion of all other means of communication by schools for the deaf. It may be augmented in all except a few who possess hardly any residual hearing by the use of hearing aids (the aural method), and is most successful in those with a high I.Q., who begin speech-training early and have a reasonable degree of residual hearing. One stage lower in the hierarchy of deaf-communication is finger-spelling. This is done by the bimanual method in this country and by the unimanual method in France and the U.S.A. Historically it antedated speech training and experts may achieve considerable speed and grammatical accuracy by this method. It has been claimed (Payne, 1918) that experts in finger-spelling are more literate than those taught by either the oral or the later aural method. Good finger-spellers are often of high I.Q. who have been neglected till late and have never acquired facility in lip-reading. The use of conventional signs is often frowned upon by good finger-spellers, good speakers, and most teachers of the deaf. But combined methods are
preferred by the majority of deaf people for signs
decrease the strain of unbroken finger-spelling or
lip-reading and add colour to their conversation.
Sign-language is ungrammatical and has yet to be
systematized. It may be, as in this patient, the sole
means of communication of those with low intelli-
gence. Confusion of meanings may frequently arise
and pass uncorrected and, particularly among the
unintelligent, lack of clarity in signing may hamper
communication.

Defects in finger-spelling due to cortical lesions,
amounting to a dysphasia for this method of com-
munication, have been noted in the past and various
explanations proposed: apraxia and agnosia for
dactylography (Critchley, 1938), finger agnosia
(Grasset’s patient, 1896), and affective disturbances
(Tureen, Smolik, and Tritt, 1951). This patient
demonstrates another cause for signing defects. His
involuntary movements have come on gradually,
and, although his spelling and literary ability have
always been poor, his family have been able to adapt
to the increasing paucity of communication result-
ing from the interference of the involuntary move-
ments. Only in the past three years have they come
to experience real difficulty. It may be surmised
that in a hearing choreic patient of similar intelli-
gence dysarthria would have hampered conversa-
tion at an even earlier stage. In fact, the chief draw-
back to communication in this patient has not been
the interruption of signing by the involuntary move-
ments but the limitation of his attention span and
instability of mood which have resulted from his
dementia.

SUMMARY

The association of Huntington’s chorea and deaf-
mutism has most probably occurred by chance.

The patient, who has always been somewhat
simple-minded and has never been able to master
any means of communication other than signing,
crude finger-spelling, and elementary reading and
writing, was nonetheless still able to be intelligible
to his family in the early stages of the chorea. His
recent breakdown in communication is almost
certainly the result of increasing dementia and not due
to the severity of his involuntary movements.

We wish to thank Dr. E. J. Ross for permission to publish
this report of a patient under his care, and Dr. Ross
and Dr. G. M. Stern for their help and advice in the
preparation of this paper.

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A deaf-mute with Huntington's chorea

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*J Neurol Neurosurg Psychiatry* 1966 29: 181-183
doi: 10.1136/jnnp.29.2.181

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