# CORTICAL DEAFNESS

#### A Case Report and Review of the Literature

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#### SUMMARY

A 48-year-old woman with triple cardiac valve prostheses became completely deaf after bitemporal embolism. The lesions were demonstrated radiologically and neurophysiological investigation confirmed intact peripheral and brainstem pathways. Experimental studies of the auditory pathways are briefly reviewed. Reported cases of cortical deafness are discussed and the rarity of the condition emphasised.

# INTRODUCTION

The primary auditory cortex of both man and animals is located in the anterior and posterior transverse temporal gyri of Heschl. Each ear has bilateral representation in the cerebral cortex and it is possible to remove the whole of the human non-dominant hemisphere without significant effect on either the pure tone audiogram or the discrimination of undistorted speech (Dandy 1933).

Considerable information in man has been derived from open brain stimulation (Penfield and Perot 1963) and open brain recording of auditory evoked potentials (Celesia 1976). Thus Penfield and Perot (1963) found that stimulation of the anterior transverse temporal gyrus elicited simple auditory sensations such as buzzing, whistling or knocking, while stimulation of surrounding areas including the posterior two thirds of the superior temporal gyrus elicited complex auditory hallucinations. Celesia (1976) confirmed the bilateral representation of the cochlea but obtained larger amplitude responses from the contralateral ear. He found a

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larger area surrounding the primary auditory cortex extending to the posterior two thirds of the superior temporal gyrus and the upper bank of the Sylvian fissure including frontal and parietal operculi where potentials of longer latency and smaller amplitude were recordable. It is known that fibres project to these areas from the medial geniculate bodies but little is known of the pathways or fate of either commissural or efferent fibres. It seems that cortical lesions which result in deafness must then be bilateral. The occurrence of such lesions in man sufficiently circumscribed to avoid gross deficit in other functions is rare.

In animals, bilateral ablation so that no cortex responsive to auditory stimuli remains and complete degeneration of the medial geniculate bodies occurs, results in inability to detect changes in the temporal pattern of tones (Diamond and Neff 1957) or change in stimulus duration (Scharlock et al. 1965) but the ability to detect the onset or changes in the frequency or intensity of sounds is retained (see Scharlock et al. 1965 for references). It should be noted, however, that there may be some difference between experimental cortical ablation and infarction in the human as the latter may also include subcortical white matter.

Auditory deficits in man resulting from purely cerebral lesions are various. The theoretical possibilities have been clarified by Earnest et al. (1977). Each case shows variable contamination with deficit in speech or other cortical function. Unilateral lesions of the temporal lobe cortex result in subtle auditory deficit. This may consist of difficulty in discrimination of distorted, interrupted or accelerated speech presented to the contralateral ear (Jerger 1960) or in localising sound in the contralateral auditory field (Sanchez-Longo and Forster 1958). By contrast, a unilateral dominant temporal lobe lesion resulting in pure word deafness may leave sound localisation quite unaffected (Gazzaniga et al. 1973). Such a lesion, which may be cortical or subcortical, is thought to isolate Wernicke's area from the ipsilateral and contralateral auditory cortex. An auditory agnosia for speech results so that the patient is unable to identify spoken words despite recognising that the sounds represent speech. An adequate neurophysiological description of such cases does not yet exist.

Pure word deafness is more commonly the result of bilateral lesions of the superior temporal cortex (Goldstein 1974). Where documentation exists, such cases usually show some degree of audiometric deficit (Jerger et al. 1972; Kanshepolsky et al. 1973). Similar bitemporal lesions may result in an agnosia for both pure tones and speech (Wohlfart et al. 1952; Lhermitte et al. 1971) or pure tones alone (Albert et al. 1972). These cases also showed some primary audiometric deficit. If, in such a case of bilateral cortical disturbance, the primary hearing deficit predominates, then true cortical deafness may be said to exist. In some cases of cortical hearing disorder, a true cortical deafness has been observed to resolve gradually into a residual agnosia for speech and other sounds with only minor residual audiometric deficit (Jerger et al. 1969; Goldstein et al. 1975). The relationship of pure word deafness (auditory agnosia) to cortical deafness may thus be regarded as relative rather than absolute. It has, in fact, been suggested that the 2 conditions represent part of a general cortical hearing disorder (Goldstein 1974).

Reports of cases with bilateral cortical lesions resulting in true deafness are rare. They begin with those of Wernicke and Friedländer (1883). This, and 2 other early reports, are reviewed by Bastian (1897). Modern neurophysiological and radiological tests were not available, confirmation of the site of lesions being obtained at post mortem examination. The first results of neurophysiological investigations in such a case were reported by Jerger et al. (1969). Radiological evidence in a second case was reported by Earnest et al. (1977). We wish to report the results of neurophysiological and radiological studies in a further case of total deafness due to bilateral temporal lobe lesions.

## CASE REPORT

Mrs. S. T., a 48-year-old right-handed white housewife presented to the cardiac department at the Middlesex Hospital in 1974 with a 6-year history of increasing dyspnoea and fatigue. She was noted to have rheumatic aortic, mitral and tricuspid valvular disease and to be in atrial fibrillation. Despite treatment with frusemide and digoxin she remained in gross congestive cardiac failure and, on 26.2.75, following cardiac catheterisation, she underwent triple valve replacement with Starr-Edwards prostheses. Post-operative recovery was uneventful and she was discharged home on 17.3.75 taking digoxin, frusemide, spironolactone and warfarin.

She was readmitted on 20.5.75 with a 2-day history of headache and confusion. She remained in atrial fibrillation but there were no signs of heart failure. She was noted to have a severe conduction dysphasia, but no other neurological signs. A technetium brain scan showed increased left temporal uptake and EEG showed excess theta and delta wave activity in the left frontotemporal region. Dipyridamole was added to her warfarin anticoagulation, which remained well controlled. At out-patient review on 16.7.75 she was noted to have a left homonymous hemianopia but the dysphasia had improved. She had reverted to sinus rhythm.

She remained well until March, 1976 when she was admitted to another hospital following 2 grand mal fits. Mild dysphasia remained but no new neurological deficit was found. The dipyridamole was discontinued but the other medication maintained.

In May 1977 she was readmitted to the same hospital following a further grand mal fit. Again, no new signs were found and the anticoagulation was well controlled. A technetium brain scan was normal and EEG again showed a left temporal excess of slow wave (2–5 Hz) activity.

A further grand mal fit occurred in December 1977, following which her husband noted that she was forgetful, being unable, for example, to prepare meals reliably. Then, on 4.1.78, while alone in a room, she was heard to cry out and shortly afterwards she was found to be totally deaf. Over the next few days she became increasingly agitated and violent, refusing to consult the family doctor. She remained deaf and her increasingly severe behavioural disturbance with paranoid ideas necessitated her admission to a local psychiatric hospital on 14.2.78, from where she was transferred to the Middlesex Hospital on 20.2.78.

She had received no anticonvulsants nor any ototoxic drugs. There was no family history of deafness with the exception of a maternal grandmother who had developed deafness at the age of 20 and of whom no details were available. There was no past history of head injury, of occupational noise exposure or of psychiatric disturbance.

On admission she was afebrile. The blood pressure was 140/90 and she was in sinus rhythm. There were signs of normal prosthetic valve function and no signs of heart failure.

General neurological examination revealed a left homonymous hemianopia to menace with normal optokinetic responses to a hand held drum. Apart from deafness, there were no other cranial nerve abnormalities. Her gait was normal and there was no disturbance of tone, power or voluntary movement in the limbs. She was able to execute skilled tasks such as peeling an orange without difficulty. The tendon reflexes were symmetrical and both plantar responses were flexor. The sensory examination was normal, there was no astereognosis or tactile inattention and graphaesthesia was normal.

She was orientated in place but, although able to state the day of the week, was unable to name the month or year. She would sit quietly but at times appeared agitated and perplexed, spontaneously uttering comments such as "what are they saying?" or "did she say something?". At times she showed paranoid ideas referring to bribery and attemped physical or sexual assault. She appeared totally deaf with no startle response to loud sounds. Later, as she became less agitated, she appeared to become aware that she was deaf and was distressed by this.

Her speech was of normal loudness but somewhat monotonous. It was fluent but showed occasional neologisms, paraphasias, and inappropriate and ungrammatical word usage. There were frequent dysphasic errors in naming common objects presented visually or by touch, e.g. "bricket" for button and "nickin pin" for scissors. She was able to read but had great difficulty with words of irregular spelling which she read phonetically. She could understand and execute simple written commands but was unable to understand more complex sentences. She could understand gestures and also use them for communication but the amount of kinetic and mimetic play was limited and restrained.

Writing was performed hesitantly only with much persuasion and showed spelling errors with preservation of script both in spontaneous writing and in copying. She could correctly add and subtract single figures but would not attempt more complex calculations. Graphical constructional ability was not testable because of apparent failure to understand the tasks. Kohs' blocks were assembled slowly but correctly. There were errors in naming colours.

During all these tests her performance was contaminated by quite marked perseveration.

# Psychological assessment

Psychological assessment (Dr. E.K. Warrington, The National Hospital, Queen Square, London) confirmed a global dysphasia in which expressive speech functions were most impaired. The patient succeeded in naming 9/25 of the Oldfield pictures, the errors consisting of both semantic approximations and literal paraphasias (e.g. Hat-Shoe, Horseshoe-Horse Cue). She scored 0/12 on Set 1 of the Advanced Progressive Matrices, and did not appear to grasp what was required of her. On the WAIS test of Block Design her score was well within the defective range, even though she did not appear to have any primary spatial loss. It thus seemed likely that there was also a moderately severe degree of generalised intellectual impairment.

## **Investigations**

The haemoglobin was 14.0 g/dl with normal platelet and differential white cell count. The ESR was 20 mm/h. VDRL and TPHA were negative. Prothrombin time was 34 s (control 12 s). Random blood glucose 4.4 mmol/l. MSU and blood cultures were sterile. Serum urea, electrolytes, calcium, proteins, cholesterol, alkaline phosphatase and aspartate transaminase were all normal. ANF was not detected. X-rays of chest and skull were normal. The ECG showed sinus rhythm with widespread ST-segment depression and T-wave inversion.

The EEG showed, in addition to the previously noted left-sided slow wave abnormality, a right temporal excess of delta and theta activity with sharp waves. Visual evoked responses were of normal latency. There was no psychogalvanic skin response or attenuation of the EEG to sudden loud noise, but normal psychogalvanic skin responses were obtained to gasp and visual menace.

Transmission CAT scan of the brain (Fig. 1) showed discrete area of low attenuation in both temporoparietal regions and in the right occipital region.

## Neuro-otological assessment

Methods used were:

- (1) Pure tone audiometry.
- (2) Delayed speech feedback.
- (3) Electric response audiometry (ERA):

(a) Brainstem electric responses (BSER): a series of 5 vertex-negative potentials with a latency within 10 ms can be recorded in response to wide-band click stimuli (Sohmer and Feinmesser 1967; Jewitt and Williston 1971; Lev and Sohmer 1972). These potentials arise from the brainstem and it has recently been suggested (Starr and Achor, personal communication) that they originate in the ascending fibre tracts of the cochleocortical pathway from cochlear nerve to inferior colliculi.

(b) Middle latency response (Geisler 1960; Mendel and Goldstein 1969): the most constant normal feature is a vertex-positive wave whose latency is 36 ms. It has been the subject of controversy since it can be difficult to separate this neurogenic potential from myogenic reflex responses, and it may be hard to detect in some normal subjects. It is thought to arise from the primary auditory projection area, although there may be an earlier component from the thalamus.

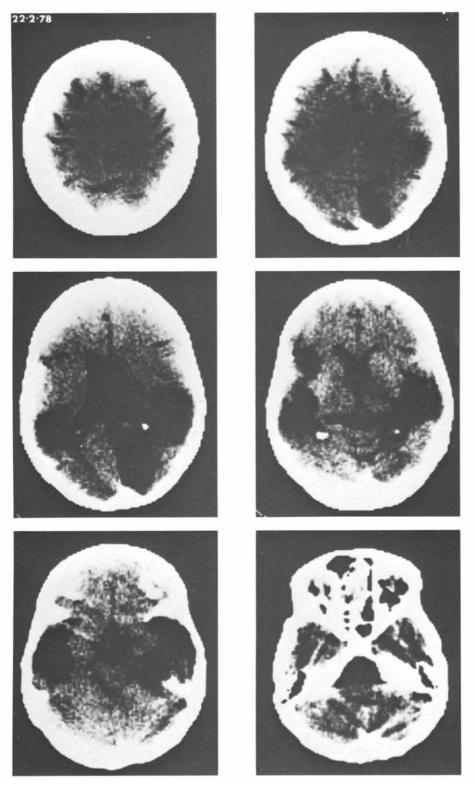


Fig. 1. Transmission computer-assisted tomographic scan showing bitemporal and right occipital lesione of low attenuation.

(c) Cortical ERA: this uses the slow vertex response or V-potential (Davis 1939; Gastaut 1953), a series of waves recorded over the vertex. The latency is 50-150 ms and their origin is the temporal lobe.

(4) Stapedius reflex: this is detected using the acoustic impedance bridge. Bilateral contraction of the stapedius muscles occurs in response to a sound 70-90 dB above subjective threshold.

# Results

Assessment was performed during the first week of March 1978.

Pure tone audiometry showed no response from either ear up to 110 dB hearing level (HL).

Delayed speech feedback testing showed no positive response using a feedback delay of 0.2 s.

The BSER using wide-band click stimuli showed normal waveforms of normal latency but with slight elevation of threshold at 45 dB HL bilaterally (Fig. 2). This probably represents a minor pre-existing peripheral lesion and is consistent with the patient's age. Cortical ERA showed no response from either ear using pure tone bursts of 120 dB HL at frequencies of 500–400 Hz. Middle latency responses were absent. Stapedius reflexes were present bilaterally although at slightly elevated thresholds.

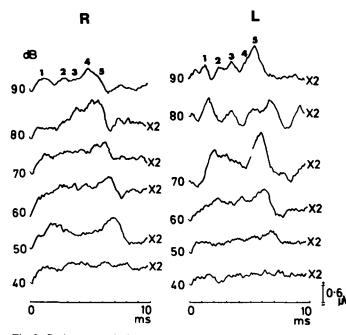


Fig. 2. Brainstem-evoked responses to 1024 averaged wide-band clicks at 40-90 dB. Wave 5 is clearly shown and is detectable down to 50 dB. It is absent at 40 dB.

### Progress

During her admission she became less agitated and her paranoid ideas subsided. She remained totally deaf. She was treated with sodium valproate and trifluoperazine in addition to her pre-existing medication of digoxin and frusemide. Warfarin and dipyridamole were continued and she was discharged home on 23.3.78.

She was admitted for reassessment on 23.5.78. Her husband reported no recovery of hearing, although her mental state had become more tranquil. There had occurred no further episodes of agitation or delusional thinking. She would spend most of her day at home sitting quietly, often reading magazines or novelettes, but her husband felt that she understood or retained little of the material read.

The cardiac signs were unaltered. She remained totally deaf. There had been no change in her dysphasia. There was evidence of a mild left sided visual inattention but no other focal neurological signs were found.

Her anticoagulation remained well controlled and routine haematological and biochemical tests remained normal. Transmission CAT head scan showed no significant change.

No response was obtained with pure tone audiometry. Cortical evoked response audiometry gave no response to 120 dB tone bursts at 500, 1,000 and 2,000 Hz in either ear. Brain-stem electric responses to wide-band clicks gave waveforms of normal latencies with, as before, slightly elevated thresholds of 45 dB in the left ear and 55 dB in the right ear.

In August 1978, she developed tremor affecting all 4 limbs. The following month she was admitted to another hospital, where she was felt to have developed phenothiazine-induced Parkinsonism which was successfully treated by orphenadrine and the withdrawal of trifluoperazine, only to become worse again when chlorpromazine was substituted because of behavioural problems.

She was reviewed again on 16.10.78. Her husband reported that at times she was able to perceive loud noises, for example, looking up when the dog barked. Most of the time, however, she seemed totally deaf and at no stage could she understand speech.

Examination showed her to be agitated with some pressure of speech and, again, evidence of delusional ideas of persecution. Her dysphasia, dysgraphia, dyslexia and visual field defect were unchanged. There was still no clinical response to the loudest sounds. There was mild intermittent rest tremor of all four limbs associated with mild rigidity. The chlorpromazine was discontinued.

## DISCUSSION

This patient is thought to have suffered 3 separate embolic lesions of cardiac origin, the first to the left temporal cortex producing dysphasia, the second to the right occipital cortex causing a left homonymous hemianopia and the third, to the right temporal lobe, resulting in total deafness. The first and third events represent left and right middle cerebral artery embolism and the second, right posterior cerebral artery embolism. Her 3 prosthetic valves continued to function satisfactorily and it is possible that these disabling cerebral infarcts might have been prevented by the permanent addition of dipyramidole to her warfarin anticoagulation (Sullivan et al. 1971).

For the first few weeks of deafness, she was agitated and paranoid. Indeed the behavioural disturbance was so severe that admission to a psychiatric hospital was required. This disturbance developed a few days after the deafness and partially resolved after seven weeks. It was thought initially to be a reaction to sensory deprivation and that the improvement coincided with increasing insight into her disability. However, in previously reported cases of cortical deafness (Table 1), behavioural problems of this type have not been mentioned, even when the deafness was complete (Earnest et al. 1977). Although the psychiatric effects of temporal lobe infarction are not prominent (Williams 1969), when agitated delirium does occur, infarction is found to be inferior and medial, in the territory of the posterior cerebral artery, and to include occipital infarction (Horenstein et al. 1967; Medina et al. 1977). As in this case, agitation tends to begin a few days after the lesion and to resolve gradually. Our patient had sustained a right occipital infarct in 1975. It is likely that the inferior and medial areas of the right temporal lobe were ischaemic prior to the right middle cerebral artery embolus in 1978 and that the latter event caused further inferior and medial temporal lobe ischaemia, in addition to lateral and superior temporal lobe infarction, and caused her psychiatric disturbance.

The cases of auditory disorder resulting from bilateral hemisphere lesions in the literature vary considerably in the stringency of their assessment. Only those reported in the last 30 years were assessed audiologically. As already noted, the deficit in most patients with a cortical hearing disorder has been agnosic. The patient is unable to recognise spoken words, and perhaps also music and other sounds, and may describe the disability with such words as "I can hear you talking, but I can't translate it" (Jerger et al. 1972; Kanshepolsky et al. 1973). The primary hearing deficit may be detected only on audiometry. Thus the patient reported by Jerger et al. (1972), who at no time complained of frank deafness, had a bilateral 30–40 dB loss for pure tones. Speech audiometry using monosyllabic words showed zero discrimination. He did, however, retain the ability to identify short sentences (60% discrimination in one ear, 30% in the other) but this ability was lost if any distortion was introduced. Similarly, Kanshepolsky et al. (1973) found a bilateral increase in pure tone threshold of 10-30 dB, impaired loudness discrimination and impairment of threshold-duration functions in their patient. However, modern techniques have convincingly demonstrated that in a few cases a major primary audiometric deficit does indeed exist. Such patients complain of being unable to hear anything, including their own speech, and may thus be said to have true cortical deafness.

Only 12 cases of significant deafness due to purely cerebral pathology have been reported (Table 1), the majority before the introduction of modern audiology. Early reports relied on post mortem confirmation of the lesions. The cases of Wernicke and Friedländer (1883), Mills (1891), Pick (1892), Mott (1907), Bramwell

Authors	Age ()	Age (yr)/Sex	Clinical events	Fathology	Comment on deafness
1. Wernicke and Friedländer (1883)	43	Ц	<ol> <li>Rt. hemiparesis, aphasia, word deafness</li> <li>Lt. hemiparesis, total deafness</li> </ol>	<sup>a</sup> Bitemporal infarction ?cause	Total and permanent (1 month)
2. Mills (1891)	46	<u>іг.                                    </u>	<ol> <li>Word deafness</li> <li>Lt. hemiparesis and partial deafness</li> <li>Non-specific illness and total deafness</li> </ol>	<sup>a</sup> Bitemporal infarction valvular heart disease	Total and permanent (9 years) quotes 3 other cases
3. Pick (1892)	24	Σ	<ol> <li>Rt. cerebral lesion</li> <li>Lt. cerebral lesion with partial and word deafness</li> </ol>	<sup>a</sup> Bitemporal infarction	Partial
4. Mott (1907)	25	ĹĹ.	<ol> <li>Transient aphasia</li> <li>Rt. hemiparesis and aphasia</li> <li>Paresis Rt. arm. nystagmus and deafness</li> </ol>	<sup>a</sup> Bitemporal emboli. Valvular heart disease	Total and permanent (7 years) but brainstem signs
5. Bramwell (1927)	62	F (R)	1. Aphasia 2. Deafness	<sup>a</sup> Bitemporal infarction. Mitral stenosis	Total and permanent (1 month)
6. Misch (1928)	29	X	<ol> <li>Endocarditis</li> <li>Dysphasia, Rt. hemiparesis</li> <li>Total deafness</li> </ol>	<sup>a</sup> Lt. cerebral haem. Haem. infarct. Rt. trans. temp. gyrus	Total and permanent (2 weeks)
7. Clark and Russell (1938)	44	F (R)	<ol> <li>Lt. hemiparesis</li> <li>Deafness; bilat. extensor plantar response</li> <li>Fatal pontine haemorrhage</li> </ol>	<sup>a</sup> Bilat. infarction of external capsule. Hypertension	Initially total, some recovery
8. Chavany (1945)	51	[L]	<ol> <li>Rt. hemiparesis, dysphasia</li> <li>Lt. hemiparesis, deafness</li> </ol>	Hypertension	Total. some recovery
9. Jerger et al. (1969)	20	M (amb)	<ol> <li>Rt. hemiparesis, dysphasia</li> <li>Complete deafness</li> </ol>	Bilat. distal middle cerebral a. occlusion on angiogram, ?cause	6085 dB loss. some recovery
10. Goldstein et al. (1975)	31	F (L)	<ol> <li>Complete deafness, Lt. hom. hemianopia, ext. plantar response, amnesia etc.</li> </ol>	Encephalitis	Total deafness evolving to word deafness over 2 years
11. Earnest et al. (1977)	27	M (L)	<ol> <li>Dysarthria, paresis Lt. face and arm</li> <li>Aphasia, Rt. hemiparesis</li> </ol>	Bitemporal infarction on CAT scan, valvular heart disease	Total deafness, recovery to 65-80 dB loss
12. Zöllner et al. (1977)	50	(T)	<ol> <li>Transient aphasia</li> <li>Deafness, Lt. hemiparesis</li> </ol>	Bitemporal lesions probably infarction	Few details. Marked audiometric loss.

TABLE I

(1927), and Misch (1928) demonstrated bitemporal infarction, with haemorrhage in the last-mentioned case, involving at least the superior temporal gyri together with varying extents of surrounding insular, parietal and temporal cortex. Where the cause is given these lesions were due to cardiac emboli. The case of Clark and Russell (1938) provides an exception, the responsible lesions being bilateral external capsular infarctions in a hypertensive. The resulting interruption of the auditory radiations, the postulated cause of the deafness, was demonstrated by degeneration of the right medial geniculate body, the left having been disrupted by terminal brainstem haemorrhage. Although the cerebral lesions appear to be sufficient to cause the deafness in these cases, in none was the state of the brainstem or peripheral pathways reported.

Further single case reports of clinical cortical deafness followed from Lemoyne (1944) and Chavany (1945). The former case, briefly initially reported by Thiébaut et al. (1944), was a 50-year-old woman whose deafness followed an episode of loss of consciousness and epistaxis. There was mild dyscalculia but no other localising signs. She was said to have an auditory agnosia and conductive hearing loss of 60–70 dB. There was no post-mortem examination. The cause of the deafness is obscure, but we suggest that it could have arisen from blood in the middle ear. Chavany's case, a 52-year-old hypertensive woman, suddenly became deaf prior to the onset of a left hemiparesis having 18 years before recovered from a sudden episode of dysphasia and right hemiparesis. Audiometry revealed significant sensorineural deafness. Although there were no reported brainstem signs, post-mortem examination was not performed, and although likely to be cortical, the cause of the deafness remains in doubt.

In 1952, Ziegler reported a 45-year-old man as a case of word deafness. The patient was seen initially 10 days after an episode of numbness of both hands, especially the right, accompanied by complete deafness. Four years before he had suffered a brief episode of reading difficulty. At the time of examination his deafness had resolved, word deafness and minor speech deficit remaining. No post-mortem examination was carried out, but a cortical aetiology of the initial deafness is possible. Lemoyne and Mahoudeau (1959) reported a case of "agnosie auditive pure avec surdité cortical". However, although the patient had "l'impression d'être devenu sourd subitement", the authors went on to state that "en réalité, il entend un léger claquement des doigts, le bruit fait par un trousseau de clefs, un léger sifflement". In addition, audiometry showed no significant deficit. This would not seem to be a case of true cortical deafness. A further case of cortical hearing loss was reported in 1963 by Hansen and Reske-Nielsen, a patient with a right hemisphere glioblastoma and cerebral oedema. The hearing loss, however, consisted of a minor fluctuating audiometric deficit rather than clinical deafness.

The report of Jerger et al. (1969) finally provided documentation of unequivocal deafness due to clearly demonstrated cortical lesions with no evidence of a peripheral lesion. They reported audiological findings in a 20-year-old man with bilateral distal middle cerebral artery occlusion of uncertain aetiology. The initial left lesion resulted in a moderate expressive and receptive dysphasia with questionable right facial

weakness and mild sensory signs in the right arm. Two months later the signs had resolved except for mild dysphasia. The right-sided occlusion occurred 7 months later and resulted in the sudden onset of deafness, apparently total, and without additional deficit. Although the patient was unable to hear speech and other sounds and did not respond to "relatively loud noises", audiometry on the day of admission demonstrated a bilateral sensorineural pure tone loss of only 60–85 dB. Shortly after admission, electrodermal audiometry showed no response at any frequency even at high sound levels. Over the next 3 months considerable improvement in pure tone audiometry occurred although marked loss at high frequencies in the right ear persisted. Speech audiometry at 3 months showed 30% discrimination in the left ear, opposite the less severely damaged cortex, and zero discrimination in the right ear. Cortical ERA was performed and no responses were obtained although the patient was able to hear the pure tone signal. No evidence of a peripheral auditory lesion was found and the absence of brainstem involvement was argued on clinical grounds.

Since 1969, there have been 4 further reports of interest. A patient with bilateral temporal lobe infarcts reported by Jerger et al. (1972) did not complain of deafness, rather reporting "I can hear you talking, but I can't translate it" and showed only a 20-40 dB loss on pure tone audiometry. The patient reported by Goldstein et al. (1975) was a 31-year-old woman with an encephalitic illness in whom there was no evidence of labyrinthine or brainstem disease and whose deafness was out of all proportion to other cortical deficit. Initially loud noises produced no response but after the first day she began to turn her head towards loud sounds although denying hearing them. Her hearing gradually improved over a period of 2 years until she could react to environmental noise and identify nonverbal sounds but remained severely impaired in her recognition of speech and music. Earnest et al. (1977) reported the case of a 27-year-old left-handed man with a Starr-Edwards valve prosthesis who presented with complete deafness following a right middle cerebral artery embolus. Four years previously he had suffered a left middle cerebral artery embolus. The lesions were demonstrated on computerised tomography (CAT scan). Immediately after the onset of deafness, pure tone audiometry showed no response while static compliance, tympanogram and stapedius reflex testing were normal. Cortical ERA performed three months later gave thresholds of 40-70 dB in each ear. Three years later, pure tone audiometry showed variable thresholds which were never less than 65-80 dB and he remained unable consistently to identify the nature, or even the presence, of sounds. Zöllner et al. (1977) described a patient with bitemporal lesions, deafness (of unspecified severity). marked pure tone audiometric loss, delayed and diminished cortical ERA (possibly a response to vibration rather than auditory stimuli) and brainstem responses of normal amplitude and latency, although of marginally elevated threshold. This case almost certainly represents true cortical deafness.

The history of our patient is similar to that of the patient reported by Earnest et al. (1977). Both patients were completely unresponsive to the loudest noises and to pure tone audiometry. Our patient showed no response to delayed speech feedback and no psychogalvanic skin response or attenuation of EEG to sudden loud sounds. It is of interest to note the absence of any startle response in these patients since the startle reaction to acoustic stimuli is well known to occur in decorticate primates and human anencephalics (see Larsson 1956). It is also apparent that purely cortical lesions may result in total deafness, despite the statement to the contrary by Howe and Miller (1975) and evidence from previously noted animal work indicating that sound may be detected at subcortical levels.

The patient reported by Earnest et al. (1977) showed only slight recovery at three years; our case has shown no improvement after nine months. The statements by Jerger et al. (1972) and DiCarlo et al. (1962) that substantial hearing loss is not a permanent consequence of bitemporal lesions must, therefore, be questioned.

Our patient showed normal stapedius reflexes and brainstem-evoked responses, effectively demonstrating the normality of the peripheral auditory pathways as high as the inferior colliculi. We believe our patient is the first with complete cortical deafness in whom brainstem-evoked responses have been reported. The absence of cortical ERA response is taken as confirming the cortical level of the auditory pathway lesion. However, it is noted with some caution that the patient of Jerger et al. (1969) showed no response on cortical ERA although able to hear the test sound, whereas the patient of Earnest et al. (1977) showed responses, albeit at elevated thresholds, but was unable to hear the stimulus.

Complete cortical deafness is therefore a most unusual event but is unquestionable as an entity. Our case illustrates nicely the use of modern audiological techniques in determining the level of a lesion in the auditory pathway.

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