A CASE OF PARTIAL DEAFNESS SIMULATING CONGENITAL AUDITORY IMPERCEPTION.

By

E. MILDRED CREAK, LONDON.

THE problem presented by the case here reported was that of inability to use, utter or comprehend ordinary speech in a child clearly not mentally defective and apparently not deaf. Such cases being comparatively rare in an ordinary children's out-patient department, it was thought that it would be of interest to produce a series of tests which could be compared with those given by Worster-Drought and Allen^{1,2} to their case of congenital auditory imperception, a detailed account of which appeared in the *Journal of Neurology and Psychopathology*. Taking this report as a model, Head's³ tests for use in cases of aphasia were given, and a comparison of the results is made later in the paper.

PERSONAL CASE.

The patient Ada W. is a girl who first attended the Children's Out-patient Department at King's College Hospital in January, 1931. She was then six years and ten months old and although lively and intelligent, and not apparently deaf, had never learnt to speak.

Family History.—She is the seventh of eight children. The mother was over 40 at the time of her birth, and labour was prolonged, due to atony. Non-instrumental delivery was effected, however, and the child cried normally. It is of interest to note that the younger brother, two years junior to the patient and the last of the family, is an intelligent child with a congenital right hemiplegia. There is no abnormality, either nervous or mental, in other members of the family or in near relatives.

Early History.—As a baby she 'took notice' in what was regarded as a normal fashion, turned her head when others entered the room, and walked when one year old. She is said to have learnt to say 'me' at ten months, but thereafter her speech failed to develop. She was easily trained in all toilet habits, and was otherwise a normal infant.

Past Health.—The usual fall on the head at eighteen months is reported but there is no history of any definite injury then or at a subsequent date. She has had bronchitis (two and a half years), whooping-cough (four years), diphtheria (five years). Measles at one year old is reported, but it was a mild attack and there were no aural or other complications.

When first seen the only complaint made was of dumbness. The child never spoke, but made several characteristic noises and sounds which she used as a language of her own. They were, however, not nearly so abundant as a corresponding vocabulary of a child of five would be. Her use of gesture was

quick and singularly vivid, and she turned her head when sounds were made behind her back. She appeared to hear the sound of her own voice, and made no grunts or spontaneous noises of which she seemed unaware. Her movements were quick and neat and there was no shuffling of the feet in walking. Her mother reported that she varied in her response to sounds: if she were busily occupied with something else she would not notice or appear to respond to a sound stimulus, but if she were listening or not so preoccupied, she appeared to hear all ordinary noises. Like the case reported by Worster-Drought and Allen, however, she appeared much less interested in sounds than in any other form of stimulus. Her powers of noticing and interpreting gesture were abnormally quick, and it was noticed that even at the age of five she watched intently the lips of those speaking to her. Her behaviour with other children was normal, and six months away from home with relatives had made no difference; she had then taken quite kindly to a family of young cousins.

On examination it was found that her tonsils were enlarged and septic and that adenoids were present. Although there was no evidence of middle ear disease, no detailed investigation was undertaken until these had been removed by operation.

She was seen again in February, 1931, three weeks after operation. She was small for her age, but her physical condition appeared to be excellent. It was thought that her hearing in the right ear was not so good as the left, but it was difficult to be sure of her replies. She was clearly not grossly deaf in either ear. All other cranial nerves were normal, and all reflexes were normal. There was no nystagmus, no incoordination, and tests for vestibular function gave a normal result.

Her voice was somewhat hoarse, possibly from disuse. Her articulation was indistinct, and her vocabulary limited to a few simple words such as 'Mummy,' 'pussy,' 'yes,' 'no,' 'no more,' and her name 'Ada.' These were quite intelligible, but she used no sentences and expressed herself without words by means of gesture. At school (ordinary elementary school) which she had attended since the age of five, it was reported that she would copy letters, but could neither read, nor write to dictation. She had learnt to count up to eight, but could not manipulate numbers in simple sums. To a Burt revision of Binet Simon test, she gave a normal response with an intelligence quotient of 95.

Clearly then, the speech defect could not be attributed to retarded mental development, and a functional mutism of such severity and consistency would hardly be postulated. In addition there was no trace of hysterical behaviour in the child.

The question of deafness had clearly to be eliminated and the difficulty of this task in a young child, inattentive to and uninterested in aural stimuli, can hardly be exaggerated.

On examination the external ear was normal, the drums were intact and unscarred, but somewhat retracted on both sides. She appeared to hear a tuning-fork (C 2) in both ears, by air and bone conduction, but it was noted that there was some loss in the right ear as the intensity diminished. This was not definite enough for a comparison with a normal ear. She turned her head rapidly to all sounds such as knocking, clapping hands or dropping objects behind her back. If her name was spoken she looked up. There was no question, however, that she understood commands more easily if the speaker's lips were visible. All vestibular tests as noted above were normal, and she showed none of the customary behaviour traits of the deaf child such as shuffling feet in walking. A provisional diagnosis of congenital auditory imperception was therefore made.

In July, 1931, arrangements were made for her to be transferred to a residential school for the deaf as it was felt that her knowledge would have to be gained by other channels than her ears, even though her capacity to hear appeared good.

In January, 1932, she was seen again, and although only seven years and ten months old, she appeared so lively and intelligent that arrangements were made to take her into hospital next school holiday and try detailed testing according to Head's scheme for aphasics. On this occasion she was seen by the aural surgeon who reported as follows: 'This child is by no means deaf. She can hear and localise loud noises. She can hear musical notes, particularly in low tones. Bone conduction to the low forks is good, and gets worse as the pitch is raised.'

In view of this report, the question was raised whether the case could be an instance of high frequency deafness. In many respects she answered to Ewing's¹ descriptions of such cases, with their 'bird-wittedness,' but without more elaborate apparatus it was agreed that such a diagnosis could only be queried. At this time (January, 1932) her mother reported that the child had learnt a good deal from school; sewing, knitting and writing when limited to copying, and drawing, were all good. She would tell the different colours and named them by signs according to a deaf-and-dumb language. She would count up to 12. The night before, at home, she had said spontaneously, 'Uncle Ted coming tomorrow,' having been told that this was so earlier in the day. But her mother observed, 'She has no sentences in her, and if she sings, it's all gabble.' Her hearing appeared to be about the same; 'She'll take notice if she's not thinking of anything, but if she's busy with anything she won't hear you.' Her behaviour was noted to be somewhat self-willed and impetuous. She had cried for two hours because she had to put on black stockings for hospital.

On this occasion she was shown a number of simple objects; these she named in an idioglossic language which, however, bore in each instance a recognizable relationship to the proper name—for example, pencil was 'betsu.' She played happily with pencil and paper and drew myself and her mother, putting in distinguishing features with great glee and commendable aptness. She drew a table with cups and saucers, tried to draw milk in the jug, gave it up, and mimicked pouring it out to show what had been intended. During this time her mother made several gentle noises which were not sufficient to attract her attention. She copied a clock face and named the numbers correctly, but mispronounced her words in a way difficult to reproduce—for example, 'twelarve' for 'twelve.' These numbers she would write to dictation, in figures, and also her own name. Reading from dictation was very poor; 'cat' and 'Ada' and 'pen' were all she could read. Results were sufficiently good, however, to indicate that more might be learnt from applying a more detailed test.

Accordingly she was seen again in April, 1932. Another intelligence test consisting of performance tests gave a result indicating an intelligence slightly above normal. Her impetuous haste and lively interest in the material sometimes spoilt her result because of premature replies and the difficulty experienced in explaining the tests with gesture only.

The following results were then obtained by applying the series of tests described by Henry Head in the investigation of aphasia. The variation introduced in their paper by Worster-Drought and Allen was used in most cases—that is, naming the object first with the observer in sight, then repeating with the observer's lips invisible to the patient. In the list of common objects, for convenience' sake, a spoon was substituted for scissors.

TABLE

TEST 1.—NAMING AND RECOGNITION

Object.	(1) Pointing to an object on the table which corresponds to one shown	(2) Pointing to an object named verbally by the observer (in sight).	(3) Pointing to an object named verb- ally by the obser- ver (out of sight).	(4) Pointing to an object named in print.
Knife	correct; rapid	correct	no response	correct
Key	" "	"	said 'bee' then chose correctly	"
Penny	" "	"	chose match	chose match
Match	" "	"	no response	" pencil
Spoon	" "	"	chose key	" key
Pencil	" "	tried knife, then key, then cor- rectly chose pencil	chose knife, then key, then laughed, said 'betsu,' chose correctly	" spoon
Key	" "	correct	chose knife	" knife
Spoon	" "	"	correct	correct
Match	" "	"	correct	chose knife
Knife	" "	"	hesitated then chose correctly	correct
Penny	" "	"	chose key, then guessed at ran- dom and further answers became valueless	said 'betsu,' then chose cor- rectly
Match	" "	"	"	chose key, then guessed at ran- dom
Spoon	" "	"	"	puzzled; no choice
Pencil	" "	"	"	chose key
Penny	" "	"	"	" match
Knife	" "	"	"	correct
Key	" "	"	"	"
Match	" "	"	"	"

1.

OF COMMON OBJECTS.

(5)	(6)	(7)	(8)
Duplicate placed in left hand (out of sight).	Naming an object indicated.	Writing the name of an object indicated.	Writing the name of an object indicated. (Test repeated on another day.)
correct; rapid	libow	Aat	AEGK
" "	ee-er	Wrtse	KOY
" "	malis (? money's)	London	GIHWT
" "	mach	Watts	KODWIHMN
" "	boo-er	Aartal	IOGHTWMNYT
" "	betsu	Yiboots	HOWg
" "	ee-er	Wrtse	KOY
" "	boot	London	IOGHTWMNYT
" "	bach	Aat	KODWIHMN
" "	lievah	Watts	GHIWT
" "	malis	Aartso	OIWT
" "	mach	Aat	OHLW
" "	bloodah	Wrtse	HEF
" "	betsu	Yiboots	REWOT
" "	maris	Wrtse	GRFE
" "	liebow	London	THG
" "	ee-er	Aat	WDRE
" "	mach-er	Aartse	OEhat

TABLE

TEST 2.—NAMING AND

Colour.	(1) Pointing to similar colour to that shown.	(2) Pointing to colour named by observer (visible).	(3) Pointing to colour named by observer (hidden).	(4) Pointing to colour named in print.
Black	correct and rapid	correct and rapid	correct and rapid	chose orange
Red	"	"	"	correct
Blue	"	"	"	"
Green	"	"	"	"
Orange	"	"	"	"
White	"	"	"	"
Violet	"	"	"	"
Yellow	"	"	"	"
Red	"	"	"	"
White	"	"	"	"
Yellow	"	"	"	"
Blue	"	"	"	chose black
Green	"	"	"	correct
Black	"	"	"	chose orange
Orange	"	"	"	correct
Violet	"	"	"	"

2.

RECOGNITION OF COLOURS.

(5) Naming colour shown.	(6) Writing name of colour indicated.		(7) Printed name read aloud.
	16.4.32.	repeated 19.4.32.	
said 'black'	blulo	balle	read 'blaa'
'whrada'	red	red	'rhée' (as in French 'crème')
'blulo'	lube	bube	'bloo-ah'
'whree-er'	green	green	'whree-er'
'orvish'	orntow	oriREtaeEF	'orvish' (and imitated sucking an orange)
'wyjo'	Wratoaln	Wattaer	'belahtay'
'valt'	AERoot	Wratt	'wahta'
'djallow'	Yellow	Yellow	'djallow'
'whrada'	Red	Red	'whraya'
'wyjo'	wratoaln	wratt	'blattaya'
'djallow'	Yellow	Yellow	'djallow'
'blulo'	lube	bube	'bloo-ah'
'whree-er'	Green	Green	'whree-ah'
'blaat'	blulo	Balle	'blatta'
'orvish'	orntow	oriREtaeEf	'orvish'
'Valt' but hesitated and did not appear to know the word	Wratoaln	Wattaer	'weissa'

TABLE

TEST 3.—THE MAN, THE

Test applied.	(1) Reading aloud.	(2) Reading from pictures.	(3) Writing from pictures.
The dog and the cat	wow-wow; alaati	correct and quick	dog cat
The man and the dog	bata; wow-wow	„	eng dog
The cat and the man	alaati; bata	„	cat eng
The cat and the dog	alaati; wow-wow	„	cat dog
The dog and the man	wow-wow; bata	„	dog eng
The man and the cat	bata; alaati	„	eng cat
	(occasionally mixed 'vee' for 'the,' but made no other attempt at repro- ducing in the form of a sentence)	(but used her own terms through- out. These the same as in pre- vious column)	

3.

CAT, AND THE DOG TEST.

(4) Writing from dictation (observer visible).	(5) Writing from dictation (observer not visible).	(6) Repetition (observer hidden).	(7) Reading what she has written.	(8) Copying.
CAT	dog	ve wow-wow	correct	correct and rapid
dog	CAT	alaati		
ACA	ACA	bata. ve	„	„ „ „
dog	dog	wow-wow		
CAT	CAT	alaati	„	„ „ „
ACA	ACA	ve vata		
CAT	dog	alaati	„	„ „ „
dog	CAT	ve wow-wow		
dog	ACA	wow-wow	„	„ „ „
ACA	dog	bata		
ACA	CAT*	ve bata	„	„ „ „
CAT	dog	alaati		
	* (heard last word clearly and wrote 'cat' at once. Then sat and puzzled and then wrote 'dog')		(Using her own terms as in pre- vious column)	

TEST 4.—THE CLOCK TEST.

The first test, that of direct imitation, was performed correctly and rapidly. On attempting the succeeding tests it became clear that the patient could not tell the time in the ordinary way, although she knew and understood the numerals (arabic but not roman) and could arrange these in order.

For instance when told to set the clock at 1.10 she tried each finger singly,

TABLE

TEST 5.—THE COIN

Command.	(1) Oral command (observer in sight).	(2a) Written command using figures, e.g., 2nd into 4th.	(2b) Written command using words, e.g., second into fourth.
Second into third	correct	Third into second	correct
First into third	„	correct	doubtful: made no response
Second into first	„	First into second	hesitated, tried third into fourth
Third into second	„	correct	tried first into third
First into fourth	„	„	Subsequent replies
Fourth into third	„	„	became quite ran- dom and it was
Second into fourth	„	„	obvious that the
Fourth into first	„	„	patient was guessing,
Third into first	„	„	i.e., she knows the
First into second	„	„	figures but not the
Third into fourth	„	„	words for numerals
Fourth into second	Second into fourth then corrected it to fourth into second		

first at 1, and then at 10. Then in all subsequent tests she decided to regard the big finger only, e.g., when given the clock set at 1.55, she called it eleven, and when given the printed command 2.55, she set the big finger at 2, and paid no attention to the position of the small finger.

The test, therefore, appeared to be valueless, except in showing that she recognized both the sight and sound of numerals.

4.

AND BOWL TEST.

(3a)	(3b)
Command read aloud by patient from figures.	Command given her to read fully written out in words.
correct	could not read it, therefore no attempt made
"	"
"	"
"	"
"	"
"	"
"	"
"	"
"	"
"	"
"	"
Her articulation lacked clearness and she said 'one-er,' 'four-er,' and 'penna' for penny. She attempted no sentences	

TABLE

TEST 6.—THE HAND,

Movement.	(1) Imitation of movement made by observer.	(2) Imitation of movements of observer seen in glass.	(3) Carrying out pictorial commands.	(4) Pictorial com- mands seen in glass.
L hand to L eye	correct	correct	*R hand to R eye	correct
R hand to R ear	„	„	*L hand to L ear	„
R hand to L eye	„ (slight move- ment to R eye checked)	„	*L hand to R eye	L hand to R eye
L hand to R eye	R hand to L eye	„	*R hand to L eye	correct
L hand to L ear	correct	R hand to R ear	*L hand to R eye	„
R hand to R eye	„	correct	correct	„
L hand to R ear	„	„	„	„
R hand to L ear	„	„	„	„
L hand to L eye	„	„	„	„
R hand to R ear	hesitated, then correct	„	„	„
R hand to L eye	correct	„	*L hand to R eye	„
L hand to R eye	„	„	*R hand to L eye	„
L hand to L ear	„	„	L to (1) R eye (2) R ear	„
R hand to R eye	„	„	correct	„
L hand to R ear	„	„	„	„
R hand to L ear	„	„	L hand to R ear (*spontaneous correction followed immediately)	„

5.

EYE AND EAR TEST.

(5) Oral commands (observer visible).	(6) Printed commands not read aloud.	(7) Reading aloud and executing commands.	(8) Writing down movements made by observer.
R hand to (1) R ear (2) R eye L hand to R ear L hand to L eye	R hand to L eye R hand to R eye R hand to R eye	correct ,, ,,	Patient could make no attempt at this.
correct	L hand to L eye, followed by rapid alternation	L hand to L eye	
R hand to R ear	L hand to R ear	correct	
L hand to L eye	R hand to L eye	,,	
L hand to L ear	R hand to R ear	,,	
correct	R hand to R ear	,,	
R hand to R eye	correct	,,	
L hand to L ear	,,	R hand to L ear	
L hand to R eye	R hand to R ear	correct	
R hand to L eye	L hand to L eye, followed by R hand to R eye	,,	
R hand to (1) R eye (2) L eye (3) L ear	correct	,,	
L hand to L eye	R hand to R ear	,,	
R hand to L ear	correct	,,	
L hand to R ear	,,	,,	

TABLE 6.

TEST 7.—THE ALPHABET.

	(1) Repeating letters.	(2) Repeating letters after observer (seen).	(3) Repeating letters after observer (not seen).	(4) and (5) Writing from dictation.	(6) Reading letters.	(7) Arranging letters in order.
A	Patient has not learnt the alphabet as such, and she failed to re- peat it in order what- ever method was tried to set her going	ayt		Writing from dictation failed because she appeared not to have learnt yet to correlate sounds with symbols	no reply	Had no idea how to arrange letters in order
B		bee			„ „	
C		tee			„ „	
D		ee			„ „	
E		ee			ce-er	
F		af			no reply	
G		khee			„ „	
H		aych			sh	
I		aier	aiee		no reply	
J		chay			„ „	
K		gal	hase		ka-ka-ka	
L		el			no reply	
M		em	esj		mm	
N		en			no reply	
O		oh			„ „	
P		bee	gee		pu-pu	
Q		choo	yover		no reply	
R		ar			„ „	
S		rouse	es		sss	
T		tee			te-te	
U		yoo	yover		no reply	
V		viz			„ „	
W		dubberzoo			w-w then said twelabees	
X		az			no reply	
Y		wise			„ „	
Z		zisala	zell		„ „	

Note: In column (3) the sound is only indicated when it differed materially from those in column (2). In the blank spaces the sound made by the patient was the same with the observer seen and not seen.

CALCULATIONS.

In these the patient was asked to add the figures, and her results are shown with the corrections she made herself. She worked from left to right, and does not appear to understand the significance of the position of the numeral indicating tens or hundreds.

She does not know how to subtract or divide.

ADDITION.

1.	2				
	4				
	—				
	7	(6)			
	—				
2.	3				
	5				
	—				
	8				
	—				
3.	1	5	7		
	6	3	2		
	—				
	10	8	9	(7	8 9)
	—				
4.	2	3	7		
	1	4	5		
	—				
	3	7	11		
	—				
5.	4	3	6		
	2	8	1		
	—				
	6	11	7		
	—				

A detailed comparison of these results with those of Worster-Drought and Allen was made, the actual number of correct and incorrect replies, and the nature of the error compared in each instance. The language ability generally, especially in tests which involved reading written commands, was poorer in this case, but here the difference in age, and consequently in the length of time spent at school, may afford an explanation. Otherwise the only outstanding difference is in Test 2, Table 2 (naming and recognition of colours). My patient appeared to be more familiar with both the sounds and the appearance of the words for the common colours, and her spoken idioglossic terms are in most cases only a distorted attempt at the correct pronunciation. Both in this test and in that preceding it the words written by the patient bear no relation to the sounds she made in naming the colour, and the impression was given to the observer that the child merely wrote down at random a series of letters, having found by experience that was what was expected of her. Her intelligence was sufficient to enable her to use the

same 'word' several times for the same object, but when the test was repeated at a later date a totally different 'word,' i.e., series of letters, was supplied. This point is stressed as being of some interest and importance, and whether the same might not apply to some of Worster-Drought and Allen's results is queried, for example HSTG for 'red.' After all, if the child has no knowledge of the ordinary system of phonetics in use in our language, and has not learned to associate certain sounds with certain symbols, why should he be able to make a coherent attempt at spelling his own idioglossic tongue?

A NEW TEST.

After the completion of these tests, another was devised which aimed at investigating not only her ability to hear, but also to distinguish and recognize different qualities of sound, and to estimate her capacity to remember and associate a given sound with the object which made it. This test was applied on April 23, and repeated as detailed on May 5, 1932 (Test 8).

Six objects, each making a characteristic sound, were shown to the patient. It was ascertained by signs that she could hear each sound, and she was allowed to handle, sound and play with each one. These were:—

(1) An ordinary umpire's whistle with a fairly shrill, high note=*whistle*: (2) A small table dinner-gong, with a low booming note=*gong*: (3) An improvised rattle made from an ordinary (empty) cocoa-tin with two lumps of sugar in it=*tin*: (4) A child's toy trumpet sounding three notes in harmony=*trumpet*: (5) A small table bell of brass, with a clapper. This gave a note of moderate pitch and intensity=*bell*: (6) A toy telephone, with a handle at one side which when turned rang a small high-pitched, rather 'tinny' bell, something similar to the bell of a small alarm clock=*telephone*. This sound was the least intense of any. She was then, by means of signs, told to draw each of these objects, making sure that she recognized and identified each one with its own sound. The first drawings were made with the objects in situ, and she was highly amused at this part of the procedure.

For the first time through she had in front of her her own sheet of drawings, as it appeared a considerable feat to draw entirely from memory so complicated an object as a toy telephone or dinner gong. It was noticeable that her drawings showed a progressive simplification and omission of detail, but each one retained sufficiently characteristic features to be easily identifiable. She then sat down to a desk with paper and pencil. The objects were hidden from her behind a cloth-covered screen, and, in the order below, were sounded, each at an average, and as far as possible constant, intensity. The test consisted in getting the patient to draw the appropriate object as soon as she heard and recognized the sound made by the observer behind the screen. There was never any doubt of her ability to hear each sound stimulus, and she listened attentively, her face lighting up as she recognized it. It was clear that the louder the sound, the easier she found it to recognize, e.g., the toy telephone gave her the most trouble, this sound having both the highest pitch and least intensity.

TABLE 7.
TEST 8.—THE SOUND TEST.

Object.	First time through.	Second time through.	Nine days later.
Whistle	drew whistle, i.e., correct	correct	correct
Gong	correct	"	"
Tin	drew telephone	"	"
Trumpet	correct	"	"
Bell	"	"	"
Telephone	"	hesitated and had to have it sounded a second time	"
Gong	"	correct	"
Trumpet	drew whistle, then laughed and pointed to her original drawing of trumpet in correction	"	"
Bell	correct	"	"
Whistle	"	"	"
Gong	"	"	"
Tin	"	"	"
Telephone	drew bell	"	"
Trumpet	correct	"	"
Bell	"	"	"
Tin	"	"	"
Telephone	hesitated: drew bell	"	"
Whistle	correct	"	"

Between the first and second time through, both of which were given on the same day, she was allowed practice with the sounds she seemed to find most difficult, either because of similarity or low intensity. These were the telephone and the bell, i.e., telephone was sounded and patient drew bell. Bell was then sounded, and again patient drew bell. She was then shown objects again, and tried again, but repeated her mistake and seemed to be getting a little fatigued or bored. She was then given the sound of the tin and drew gong. The screen was then withdrawn, so that she could see all the objects, but still she pointed to the gong. She was then made to sound it, and was comically surprised at the 'wrong' sound, and immediately, without any further sounding, she picked out correctly the tin. When the test was repeated at a later date, the utmost care was taken that no sound should be made by any of the objects while exposed to view. But in order to refresh her visual memory they were shown to her grouped on a tray. They were then again hidden by the same screen, and sounded through in the order as before, and she drew, each time correctly, the object corresponding to the sound made.

It would be of great interest to repeat this test with a genuinely aphasic child, as it appears to afford a better method of estimating the patient's ability to associate sound with a variety of 'meaning' than do words in a

child who has not learnt to use them, although the test appears clearly to be one of memory rather than comprehension. A somewhat similar test was applied by Morison⁷ to his case, the auditory acuity for sounds being tested by counting the number of soft peals of a bell and the number of times the fingers were snapped. His patient recognized a mew, a bark and a low, although no details are given as to how the test was made. Although able to do all this correctly, Morison's patient made no response to words.

At the conclusion of these tests, the patient was given paper and pencil on several occasions in order to draw objects in front of her, and pictures from imagination. Her reproduction of objects was usually accurate and showed that she observed carefully any characteristic features, but on two occasions she showed a tendency to a form of mirror drawing in which the direction of converging lines, thus $\diagup \diagdown$ were reproduced $\diagdown \diagup$. This was shown in her drawings of an inkwell, and of a small calendar on a stand. Her spontaneous drawing consisted of flowers standing in front of a scribble she called 'wahtah' (? water). She then drew in dots all over this, and indicated by pointing out of the window, where it was raining, that these indicated raindrops. She described this picture by a phrase which sounded like 'evadnayow.'

Her behaviour with a piano was of some interest. She played with it, and heard the sounds made by various notes, but did not betray the usual pleasure of a child allowed to strum. She does not sing spontaneously, and on getting her to listen and copy a note played or sung by the observer on a definite vowel sound, as for example 'oo' on middle C, she would sometimes pitch it correctly but often not. Her attempts to sing simple intervals e.g., Doh-mi-soh, after hearing them played on the piano, failed at first, but she showed improvement with practice. Some evidence was thus afforded that she hears and discriminates between sounds, and that she can hear her own voice.

At this time, May, 1932, her ears and hearing were tested again, and as before the only unequivocal sign elicited was an increase of bone conduction over air conduction. The extent to which air conduction was reduced was impossible to ascertain owing to her uncertain and variable replies. Weber's test was referred to the right and her hearing appeared to be better in her left ear, although again no gross hearing loss could be demonstrated by ordinary means. Vestibular tests in a rotating chair were normal, typical nystagmus occurred and the head was held inclined to the side at the end and immediately after rotating in the chair. On recovery from the test, she was able to walk straight with open and closed eyes, and caught a ball neatly and accurately. Dr. F. L. Golla, who saw the patient and performed the tests, was satisfied that there was no impairment of vestibular function. The aural surgeon's report on April 25 gave as an opinion, that the case was one of congenital aphasia 'which had been made worse by Eustachian obstruction and resulting retraction of membranes. Hearing is better than when I last saw her, particularly does she hear the lower-pitched sounds.' Inflation was regarded as out of the question owing to the difficulty of obtaining cooperation in so young a child.

THE AUDIOMETER TEST.

While investigating this case, an opportunity occurred to have the child tested by Dr. A. W. G. Ewing, using his special adaptation of the Western Electric (2A) audiometer. The technique of this hearing test is fully described in Ewing's work on aphasia, and more briefly in a recent article in

the *British Medical Journal*⁶. The outstanding points are that a sound stimulus is given consisting of a pure tone, and that this is heard by the subject through an ear-phone applied in succession to either ear. The instrument carries a range of octaves and each note can be sounded at different intensities by means of varied resistances, thus covering the range of the human voice. The child was instructed by means of dumb show to press a button as soon as any stimulus was perceived. Her ready interest in this test demonstrated her ability to comprehend the instructions and her replies were every time consistent, a check being introduced by testing down from a clear stimulus to the point at which this became inaudible, and on the second time through, working from silence to the first perceptible sound.

AUDIOGRAM OF ADA WRIGHT, APRIL 18, 1932.

Line at 'A' indicates reading of audiometer when testing a person of normal hearing with the D-80904 conduction receiver.
L=Left ear of patient. R=Right.

The result of this test was to demonstrate convincingly the presence of a moderately severe degree of deafness, resulting in partial hearing loss over a large field (see fig.).

The chart indicates the condition graphically, and Dr. Ewing's comment was as follows:—'Audiometer test indicated 50-60 per cent. hearing loss for speech. Readings from left ear were better than right. Tests with the human voice afforded corroboratory evidence. This degree of deafness, where the trouble is of early incidence, usually results in the child making few and imperfect attempts to speak. In my opinion this severe deafness is the sole cause of the speech difficulty.'

The whole aspect of this case was, therefore, altered and it appeared merely as if a mistake in diagnosis had been made due to lack of ordinary care. On reflection however, certain points emerged which appear to be of interest, and possibly of significance in the consideration of other similar cases.

DISCUSSION.

All authorities are agreed that for young children tuning-fork and other hearing tests are highly unsatisfactory. On three occasions this child was tested by experts, and on many others observed in her reactions to sounds, and a unanimous conclusion was reached that she was not seriously deaf. That some alteration was present was agreed, and on the first occasion she was seen her enlarged and septic tonsils and adenoids were noted, together with the retraction of the tympanic membranes. But this state was not regarded as a possible explanation for the whole of her disability, and while there is some evidence to suggest that the lesion is not congenital, there was no illness or inflammatory disease noted to account for the severe deafness demonstrated by Ewing's method. The pathological nature of the condition, therefore, remains an open question.

In this child the outstanding disability was in connexion with speech, on both the motor and receptive side. This was in contradistinction to her apparent alertness in response to sounds or mere noises. Even her mother thought she could 'hear' speech because of the frequency with which the attention could be gained by a loud call. The fact that she apparently heard without understanding, unless in a position to lip-read, was the feature which first led to the investigation of the case. That she can, however, both hear and understand words with which she is familiar is shown in some of her responses, e.g., in naming colours (Test 2, Table 2, column 3). This brings up the question of distortion and its bearing on the phonetic system of language training.

Ewing and others have demonstrated, by means of phonograph records, the speech of an ordinary human voice as it sounds to the partially deaf person. Although clearly audible it is quite unintelligible owing to confusion of many of the vowel and consonant sounds. A similar experience is met with by the normal person in listening to a faulty wireless announcer, or a poorly connected 'phone call. The repeated 'I can't hear you' should really be 'I can't distinguish what you say.' The difficulty in acquiring speech results from this confusion, but it is nevertheless remarkable that a faculty so universally developed with so little conscious learning or teaching should be almost totally absent as the result of this defect alone.

A further point of interest is that the 'aphasia' resulting in this child appeared in no essential to be different from the aphasia described in their case by Worster-Drought and Allen, although there the defect was postulated

as a central defect, and here it was discovered to lie in the peripheral sense-receptor. It should be noted, however, that the tests employed were devised for the investigation of a previously normal function now destroyed, while here they are used, perhaps less legitimately and less effectively, in cases where speech has failed to be acquired. The results of the discrimination between different sounds, not related to words (Test 8), appear to have a definite bearing on this point, and it would be interesting to repeat a similar test on genuinely aphasic children, that is to say, on cases of congenital auditory imperception.

In comparing and contrasting this case with others of congenital aphasia, idioglossia and congenital auditory imperception reported in the literature, certain points appear to be outstanding. Firstly with regard to the question of deafness, in nearly every case some note is made to exclude this, but little or no account is given of actual tests, except in the case of Worster-Drought and Allen. Their case was examined in detail, and hearing appeared to be normal, but a possibly scarred tympanic membrane and two operations for the removal of adenoids in early life are noted. At three, their patient had a fall after which there was 'a discharge of watery fluid and blood' from one ear. These authors also note that 'there is a certain lack of appreciation of the exact nature of the sounds he hears, as he occasionally repeats the words imperfectly, and at times fails to repeat musical notes correctly, though at other times he can do so.' McCall's⁷ case was described as a mouth-breather who had adenoids, and a peculiarly intoned speech. The child depended on lip-reading and was noted to turn towards a noise, rather than in response to speech. Bronner⁸ comments on this case and two others reported by Thomas⁹, also two cases reported by Town¹⁰, in all of which reasonable doubt might be thrown on the condition of the auditory fields in the early years of life. In Wilbur's¹¹ ten cases, eight have other physical or mental defects, including one with 'scrofula.'

In this connexion it is interesting to compare this case of Wilbur with that of a child of three, Pauline H., who was admitted to the Maudsley Hospital suffering from 'dumbness' following a shock. She was found to have enlarged tonsils and cervical glands, and subsequently developed a severe adenitis, requiring opening and draining, and double otorrhœa. She indicated her wishes by signs, and made no attempt to speak, and her cry had a peculiar monotonous, almost brassy intonation. It was difficult to ascertain how well she heard, but on one occasion she was seen to turn round and indicate that she had heard a local school bell ringing for the children at 9 a.m. She has since recovered completely, and is now speaking normally; but whatever hearing loss she had, it was severe enough to cut short for nearly 18 months her entire speech development, and indeed she ceased to use those words she had already learnt. This case would seem to

afford suitable material for comparison with a case reported by Drinkwater¹², in which there was a period of temporary severe deafness which, on clearing, left an inability to understand speech. This is ascribed to 'word-deafness,' but might surely also be due to residual deafness producing distortion. In another of Wilbur's cases, he speaks of the speech disability being associated with partial deafness, and in another with 'deafness in the perceptive ear.' In Macready's¹³, Kerr's¹⁴, Haddon's¹⁵, Taylor's¹⁶, and Sainsbury's¹⁷ cases, only brief reference is made to the examination of the ears and hearing.

Bronner⁸ notes the frequent association of behaviour problems with such individual defects as these in children who are otherwise not backward in general mental ability. Ada W. has never shown any antisocial behaviour, but she shows to some extent the 'bird-wittedness' and hasty impetuous temper which Ewing regards as somewhat characteristic of his cases of high frequency deafness.

The familial character of the defect is brought out by Worster-Drought and Allen, Barton Hall¹⁸ and Morison⁵, and E. B. Macready¹⁹ speaks of such cases as biological variants. In this family, there is no known difficulty in acquiring speech and no familial deafness. But if the deafness in this case were congenital, it is of interest that the only younger sibling shows a more severe defect in his congenital hemiplegia. Such associated anatomical defects and peculiarities such as left-handedness and supernumerary digits, noted by Sainsbury and Haddon, are not present here.

In those cases in which especially the idioglossia is studied, there does appear to be a certain similarity in the speech. L. Guthrie²⁰ points out, however, that all examples of idioglossia are but imperfect attempts at correct pronunciation, so that a similarity would be expected. The extreme difficulty in transcribing the speech of these children makes a more accurate analysis of their own individual words a very difficult matter. The frequent addition of a suffix '-er' in this case is very noticeable, as in 'chree-er' and 'bloo-er' for 'green' and 'blue,' and this same peculiarity is also noted in Taylor's case.

In conclusion this case indicates the degree to which crippling deafness in young children may be overlooked or underestimated when looked for, and further, that such deafness may be congenital or may at all events exist without any observed pathological cause. These children may show responses to sounds, especially loud noises, while appearing to be deaf to ordinary human speech, and this led, in the case here described, to a confusion between 'word-meaning deafness' which is regarded as a central lesion, and is the characteristic feature of true congenital auditory imperception, and a lowering of auditory acuity, which was great enough to prevent the acquisition of language by an otherwise intelligent child. It is suggested that the distortion of speech which occurs in the partially deaf is a sufficient

explanation of this feature. It is not suggested that cases of congenital auditory imperception do not occur, in which a complete failure to understand and acquire speech coexists with a perfectly normal range of hearing. Such a case is quoted by Ewing, and the results of an audiometer test are described in detail. Some cases reported in the literature, however, are suggestive of a partial hearing loss occurring in early life, and some designated 'aphasia' actually describe some degree of deafness. In this connexion the unsuitability of tuning-fork tests for young children is evident, particularly as these children are inclined to acquire the habit of not listening to sounds. Instead they learn to lip-read, and whether the defect is central or peripheral, they depend increasingly on visual memories for the acquisition of language. The use of the audiometer in preference to any form of tuning-fork is stressed by Ewing, and a recent communication from the Institute of Hygiene and Tropical Medicine²¹ suggests that this form of testing will come to be increasingly used.

SUMMARY.

A case is described of difficulty in acquiring and understanding speech in an intelligent child of eight years.

A detailed examination of language capacity was made with Head's tests for aphasia, and others were introduced to test the accuracy of auditory perception and the ability to retain auditory memories.

The results of aural examination by ordinary (tuning-fork) and special (audiometer) methods, are described, and the results contrasted.

A comparison is made with this case and others in the literature, with particular reference to that reported by Worster-Drought and Allen.

I am grateful to Dr. W. P. Sheldon, in whose clinic the case was first seen, and to Dr. Mapother, Medical Superintendent of the Maudsley Hospital, for permission to publish this case. I would like also to acknowledge the kindness of Dr. and Mrs. Ewing in making possible the audiometer tests.

REFERENCES.

- ¹ WORSTER-DROUGHT, C., and ALLEN, I. M., *Jour. Neurol. and Psychopathol.*, 1929, ix, 193, 289.
- ² id., *ibid.*, 1930, x, 193 (with full list of references).
- ³ HEAD, H., *Brain*, 1920, xliii, 87.
- ⁴ EWING, A. W. G., *Aphasia in Children*, 1930.
- ⁵ MORISON, A. G., *Jour. Neurol. and Psychopathol.*, 1931, xi, 28.
- ⁶ EWING, A. W. G., *Brit. Med. Jour.*, 1932, i, 1126.
- ⁷ MCCALL, E., *Brit. Med. Jour.*, 1911, i, 1105.
- ⁸ BRONNER, A. F., *The Psychology of Special Abilities and Disabilities*, 1919.
- ⁹ THOMAS, C. J., *Ophthalmoscope*, 1905, iii, 380.

- ¹⁰ TOWN, C. H., *Psychol. Clinic*, 1911-12, v, 167.
 - ¹¹ WILBUR, H. B., *Amer. Jour. Insan.*, 1867, xxiv, 1.
 - ¹² DRINKWATER, H., *Liverpool Med.-Chir. Jour.*, 1911, xxxi, 384.
 - ¹³ MCCREADY, E. B., *Arch. of Pediat.*, 1910, xxvii, 506.
 - ¹⁴ KERR, J., *Brit. Med. Jour.*, 1900, i, 1231.
 - ¹⁵ HADDON, W. B., *Jour. of Ment. Sci.*, 1891, xxxvii, 96.
 - ¹⁶ TAYLOR, F., *Med.-Chir. Trans.*, 1891, lxxiv, 191.
 - ¹⁷ SAINSBURY, H., *Jour. of Ment. Sci.*, 1889, xxxiv, 533.
 - ¹⁸ BARTON HALL, S., and M., *Jour. Neurol. and Psychopathol.*, 1931, xi, 304.
 - ¹⁹ MCCREADY, E. B., *Amer. Jour. Psychiat.*, 1926, vi, 267.
 - ²⁰ GUTHRIE, L., *Functional Nervous Disorders in Children*, 1907.
 - ²¹ Correction of Hearing Defects, *Brit. Med. Jour.*, 1932, i, 1139.
- Reference (but not in the text) has also been made to:—
- HEAD, H., *Aphasia and Kindred Disorders of Speech*, 1926.
- WILSON, KINNIER, *Aphasia*, 1926.